
1 Definitions

In this chapter heart failure is introduced and a brief historical outline of the condition is given before moving on to explain the terminology and classifications used to describe and define heart failure. There are discussions of the confusion of language, the impact on patients and the effects of definition on healthcare service provision.

INTRODUCTION

Heart failure is a powerful but imprecise phrase. If you asked a dozen patients or clinicians to define heart failure then you would get at least a dozen different answers. One common theme would be that heart failure is serious. Everyone understands that the heart is essential for life, so to hear it is ‘failing’ is, at the very least, worrying for patients and their carers. The phrase is imprecise because it does not explain if the heart is failing completely or partially, imminently or in the future, permanently or temporarily, what the cause is and whether a cure is possible.

Patients are often unsure what heart failure means for them. Sometimes they think that heart failure is a disease. It is not; it is a syndrome – a collection of signs and symptoms indicating certain anatomical and physiological changes. Heart failure is the end result of a number of diseases – for example, ischaemic heart disease – but is not in itself a disease. For many patients the phrase heart failure might be understood to mean the heart is about to stop. Heart failure does increase the risk of sudden death but that is not what heart failure means: the phrase is a clinical diagnosis – meaning a syndrome characterised by a reduction in the heart’s pumping function, with associated complications.

Many eminent clinicians and scholars have attempted to define heart failure more precisely (see Box 1.2). This can be done in a number of ways: for example, it can be described in terms of the structural changes in the heart muscle; or the functional changes in how the heart performs; or it can be described as either acute or chronic depending on its presentation. Whilst in some ways helpful these definitions can also hinder understanding – a patient with structural changes will also have functional changes sooner or later and a patient with acute heart failure only rarely does not also have a background of chronic heart failure, even if undiagnosed.

Box 1.1 A Basic, Functional Definition of Heart Failure

- Heart failure is the failure of the heart to supply (by a pumping action) the volume of blood required to match the demands of the body.
- This reduction in supply is due to a number of potential causes but usually is the failure of the left ventricle to contract well enough.
- The mismatch between supply and demand occurs when demand is at its highest – i.e. on exertion – however, if supply is very poor then it can occur when demand is still relatively low – i.e. at rest.

HISTORICAL OUTLINE

Discussions of heart failure rightly concentrate on current discoveries and treatments so it is sometimes forgotten that heart failure is not a new condition and that understanding of the condition has come about incrementally. An appreciation of the history of the condition is a reminder that current understanding will also be incomplete and perhaps even simply wrong in some respects – a useful point to bear in mind when patients respond differently to expectations.

There are descriptions in classical literature that fit the clinical picture of heart failure (Davis et al. 2000). The Romans were aware of the medicinal (and toxic) effects of the foxglove (*digitalis*) plant on the heart. This knowledge was passed on through folklore before rediscovery by the medical profession in 1785 by William Withering, a Birmingham physician, who published a paper on *digitalis*, having seen a woman use it for a patient suffering with *dropsy* – what we would describe as congestive cardiac failure (Bessen 1986).

From the nineteenth century through to the twentieth century the development of investigations such as X-rays, electrocardiographs, echocardiographs and cardiac catheterisations has helped our understanding of what happens when the heart fails. These developments continue with the use of magnetic resonance imaging (MRI) and computer modelling techniques.

Effective treatments have taken longer to emerge. In medieval and early modern times patients with heart failure had, like the sufferers of most conditions, blood drained. This was based on the classical belief that the body had four humours and that an excess of blood caused certain conditions (Ventura & Mehra 2005). It is difficult to imagine that this did anything but make the situation worse physiologically by leading to anaemia and a compensatory neurohormonal response. However, it was the expected treatment of the time and no doubt it had a positive placebo effect for some patients. In the nineteenth and twentieth centuries a slightly different treatment emerged – that of fluid removal from oedematous tissue by inserting drainage tubes, invented by Dr Southey and hence known as Southey Tubes, into the subcutaneous tissues (Davis et al 2000). This may have provided some local

improvements in the oedema but it was not treating the cause of the oedema and any improvement would have been only temporary.

Fluid removal continues to be a mainstay of treatment in heart failure patients with fluid retention but it is now removed chemically by drugs rather than mechanically. Diuretics, or drugs that make urine production increase, came into wide clinical practice in the 1950s and remove excess fluid from the body. In the late twentieth century a better understanding of the neurohormonal responses in heart failure led to the development and use of drug therapies to affect these: angiotensin-converting-enzyme inhibitors, angiotensin II receptor blockers and aldosterone antagonists for the renin-angiotensin-aldosterone system; beta-blockers for the adrenergic system.

From the late twentieth century surgical and technological treatments have been developed for patients with heart failure. These include pacemakers, internal defibrillators and surgical treatments. For some patients there are the options of replacing the heart through a transplant or an artificial cardiac support device.

Although heart failure has no doubt been around for as long as humans have existed, it has become a more prominent condition in recent years. This is partly due to advances made in recognition and diagnosis but mainly it is due to the fact that as people live longer it becomes more common. This is for two reasons: firstly, older people are more likely to have had primary cardiac events that may damage their hearts and have had longer exposure to cardiac risk factors; and secondly, the heart, like all organs, naturally loses efficiency and robustness as it ages.

DEFINITIONS

Patients are, not surprisingly, keen to have their condition defined – to get an answer to the simple question, ‘*What is heart failure*’? Unfortunately for clinicians it is not an easy question to answer simply. Eminent cardiologists have come up with different definitions (see Box 1.2).

Box 1.2 Expert Definitions of Heart Failure

‘A condition in which the heart fails to discharge its contents adequately.’

(Lewis 1933)

‘A pathophysiological state in which an abnormality of cardiac function is responsible for the failure of the heart to pump blood at a rate commensurate with the requirements of the metabolising tissues.’

(Braunwald 1980)

‘A clinical syndrome caused by an abnormality of the heart and recognised by a characteristic pattern of haemodynamic, renal, neural and hormonal responses.’

(Poole-Wilson 1985)

Box 1.2 (Continued)

‘Symptoms of heart failure, objective evidence of cardiac dysfunction and response to treatment directed towards heart failure.’

(Task Force of the European Society of Cardiology 1995)

‘Heart Failure is a complex syndrome that can result from any structural or functional cardiac disorder that impairs the ability of the heart to function as a pump to support a physiological circulation. The syndrome of heart failure is characterised by symptoms such as breathlessness and fatigue, and signs such as fluid retention.’

(National Institute for Clinical Excellence 2003)

Quotations from Davis et al. 2000 and NICE 2003

There are various problems with defining heart failure for patients and heart failure is an uncomfortable diagnosis for patients, clinicians and health service managers.

For patients the phrase is highly value-laden. Patients know that anything to do with the heart is serious and potentially fatal. The word ‘failure’ has a socially constructed meaning that is negative: it is the opposite of success. There is also, for some, an implication that the individual has responsibility for the failure. We all want to be successful and are often regaled with anecdotes about how our successes or failures are up to ourselves. A parallel might be drawn with cancer patients and the way in which patients can feel under societal pressure to ‘stay positive’. It is curious how this pressure seems to apply only to conditions that are either life-threatening or have limited treatments: there does not seem to be a parallel pressure to stay positive, as a determinate of the success of the treatment, when having a cataract operation.

As noted earlier, for patients the phrase is unclear – does it mean that the heart is about to stop? Patients also easily confuse heart failure, myocardial infarction and arrhythmia. This is hardly surprising as they are linked and patients could of course have them simultaneously. Psychologically, the phrase heart failure and the explanation that it is a progressive and chronic condition are difficult for the patient to deal with as people are used to stories of medical interventions and successes and expect something to be available as a cure. It is important that definitions are clear and comprehensible. This means clinicians must avoid jargon, medicalisation and over-complication. It means clinicians must be ready to adapt their explanations to suit the patient’s understanding. Communication failures are, regrettably, quite common in clinical practice – for example, in one study of heart failure 40% of patients had no understanding of the nature or seriousness of their condition (Buetow & Coster 2001b). It is sensible practice to check patients’ understanding in later consultations and to build up their knowledge gradually – see Chapter 8.

For the clinician the problem with the phrase heart failure is that it is so broad. Although heart failure is usually a functional phrase it is imprecise: a heart may be failing in the sense that it is not going to last another 24 hours; or it may be failing in the sense that a weakness has been diagnosed for many years; or even, taking the American Heart Association staging system, that there are risk factors or precipitating diseases for heart failure. It is also imprecise as a definition because the heart can fail in different ways – hence the use of more structural terms such as left-sided, right-sided, biventricular and congestive cardiac failure. It is also possible to define heart failure using descriptive terms such as high output, low output, diastolic and systolic dysfunction. Other definitions try to link to specific changes in the structure or tissues of the heart muscle – such as dilated cardiomyopathy – sometimes with the addition of the underlying cause – such as alcoholic dilated cardiomyopathy. The common criticism of these definitions is that they are reductionist. It could also be argued that defining conditions in medical terminology eases the explanation process for the clinician by medicalising the condition but excludes the patients by setting up a professional terminology barrier. The risk of telling the patient that they have *dilated cardiomyopathy* is that they might have no idea that it means they have *heart failure*. Various clinicians have advanced alternative phrases to heart failure but even the relatively minor adjustment from *heart failure* to *cardiac failure* is a move away from plain English.

For the health service the problem of the definition of heart failure is also a real one. This is because in order to tackle, analyse and resource a problem you need to know what the problem is and what it is not, how widespread it is and what resources it uses. Using a closed definition of heart failure (such as left ventricular systolic dysfunction confirmed on echocardiogram, as in the new GP contracts), a smaller number of patients will be found than if a broader definition (such as including diastolic dysfunction and those with clinical evidence but no echocardiogram confirmation) is used. Using a broader definition of heart failure may be best if the policy aim is to reduce the burden of heart failure or future presentation. Conversely, if the aim is to target resources to those with existing symptomatic heart failure, a narrower definition of heart failure is best. For health service managers a further complication with definitions is that different types of heart failure need different types of funding – a patient with genetic cardiomyopathy may not have many admissions but if they do they may be high cost; and as they are diagnosed younger than the average heart failure patient, their healthcare costs are over many more years.

TERMINOLOGY

CARDIOMYOPATHY

The term cardiomyopathy began being used in the 1950s and was first classified by the World Health Organization in 1980 (World Health Organization 1980).

Cardiomyopathy means disease of the heart muscle. The World Health Organization definition can cover the majority of cases of heart failure, the exceptions being reversible functional causes such as acute anaemia and thyrotoxicosis.

Box 1.3 WHO Cardiomyopathy Classification

Intrinsic to myocardium

- Dilated Cardiomyopathy (DCM)
- Hypertrophic Cardiomyopathy (HCM)
- Arrhythmogenic Right Ventricular Dysplasia (ARVD)
- Obliterative Cardiomyopathy (OCM)

Secondary to external processes

- Ischaemic Cardiomyopathy
- Hypertensive Cardiomyopathy
- Valvular Cardiomyopathy
- Inflammatory Cardiomyopathy
- Cardiomyopathy due to other systemic diseases.

A more detailed classification of cardiomyopathy is provided by the American Heart Association and has recently been revised (Maron et al. 2006).

Box 1.4 AHA/ACC Cardiomyopathy Classification

Primary Cardiomyopathies

Genetic	<ul style="list-style-type: none"> • Hypertrophic Cardiomyopathy • Arrhythmogenic Right Ventricular Dysplasia (ARVD) • Conduction System Disease (Lenerge Disease) • Ion Channel Diseases (Long QT Syndrome; Brugada Syndrome; Catecholaminergic Polymorphic Ventricular Tachycardia; Short QT Syndrome; Idiopathic Ventricular Fibrillation)
Mixed (Genetic and Non-genetic)	<ul style="list-style-type: none"> • Dilated Cardiomyopathy • Primary Restrictive Non-hypertrophied Cardiomyopathy
Acquired	<ul style="list-style-type: none"> • Myocarditis (Inflammatory Cardiomyopathy) • Stress ('Tako-Tsubo') Cardiomyopathy • Peripartum Cardiomyopathy • Arrhythmia Induced Cardiomyopathy • Alcoholic Cardiomyopathy

Box 1.4 (Continued)*Secondary Cardiomyopathy*

Infiltrative	<ul style="list-style-type: none"> • Amyloidosis • Gaucher Disease • Hurler's Disease • Hunter's Disease
Storage	<ul style="list-style-type: none"> • Haemochromatosis • Fabry's Disease • Glycogen Storage Disease • Niemann-Pick Disease
Toxicity	<ul style="list-style-type: none"> • Drugs • Heavy Metals • Chemicals
Endomyocardial	<ul style="list-style-type: none"> • Endomyocardial Fibrosis • Loeffler's Endocarditis
Inflammatory	<ul style="list-style-type: none"> • Sarcoidosis
Endocrine	<ul style="list-style-type: none"> • Diabetes Mellitus • Hyperthyroidism • Hypothyroidism • Hyperparathyroidism • Pheochromocytoma • Acromegaly
Cardiofacial	<ul style="list-style-type: none"> • Noonan Syndrome • Lentiginosis
Neuromuscular/Neurological	<ul style="list-style-type: none"> • Friedreich's Ataxia • Duchenne-Becker Muscular Dystrophy • Myotonic Dystrophy • Tuberous Sclerosis
Nutritional Deficiencies	<ul style="list-style-type: none"> • Beriberi (Thiamine Deficiency) • Pellagra • Scurvy • Selenium • Carnitine • Kwashiorkor
Autoimmune/Collagen	<ul style="list-style-type: none"> • Systemic Lupus Erythema • Dermatomyositis • Rheumatoid Arthritis • Scleroderma • Polyarteritis Nodosa • Electrolyte Imbalance
Post-cancer treatment	<ul style="list-style-type: none"> • Radiation • Anthracyclines • Cyclophosphamide

As can be seen from these classification systems the term cardiomyopathy can be helpful when trying to move from a clinical diagnosis based on presentation and investigation to consideration of the underlying cause of the condition. It is important to try to do this because, as the lists clearly show, there are a great many potential causes of cardiomyopathy and they have different implications for patients and their families. However, in clinical practice, patients often present with several possible causes of heart failure and it is not always possible to determine where they would fit in a classification system.

LEFT VENTRICULAR FAILURE (LVF)

Left ventricular failure is often the admission diagnosis of acutely unwell patients admitted to hospital with heart failure. Left ventricular failure is also the usual cause of chronic symptoms such as breathlessness in patients with heart failure.

The left ventricle is the heart chamber that pumps blood through the aortic valve into circulation. In order to do so it has the highest pressure and largest muscle bulk of any of the heart chambers. There are several ways in which the left ventricle can fail. It may fail either acutely or chronically. It can be too baggy (dilated), or the chamber size might be too small because the muscle is over-thickened (hypertrophy). There might be problems with the ability of the muscle to contract – either the power or the co-ordination of the contractions.

The most common left ventricular problem in developed countries is as a consequence (either immediate or longer term) of ischaemic heart disease. The workload of the left ventricle creates a high demand for blood. The left ventricle has the largest coronary blood supply, making it the most likely to be involved in myocardial infarctions. Damage to the left ventricular myocardium through infarction leads to reduced contractility and abnormal movement of the walls. The result is either reduced cardiac output or a need for the ventricle to increase its workload to maintain the same cardiac output. This in turn leads to an increase in muscle bulk (hypertrophy) and an enlargement of the chamber (dilation).

These physical changes in the myocardium are known as remodelling and are mediated by the effects of hormones. Although a compensatory mechanism the remodelling of the heart has a negative effect eventually as the ventricle, in effect, becomes ‘over-stretched’ and the fibres lose their elasticity. This is discussed more in Chapter 5.

RIGHT VENTRICULAR FAILURE (RVF)

Although the left ventricle is the most common ventricle to fail (and some people mistakenly assume left ventricular failure and heart failure are synonymous) it is possible that the right ventricle can fail without the initial involvement of the left ventricle.

This can occur in right ventricular myocardial infarctions or in patients who have severe lung disease. Patients who develop right-sided heart failure as a secondary result of lung disease are said to have *cor pulmonale*.

Right ventricular failure presents differently to left ventricular failure and this will be discussed in Chapter 6. Eventually, a failing right ventricle will lead to a failing left ventricle just as a failing left ventricle eventually leads to a failing right ventricle.

BIVENTRICULAR FAILURE OR CONGESTIVE CARDIAC FAILURE (CCF)

This is the term used when both ventricles are failing simultaneously. As you would expect, biventricular failure is suggestive of a more advanced heart failure, with more complications and worse outcomes. The term congestive cardiac failure was used in order to describe the pattern of fluid retention and pulmonary oedema usually present. It is a phrase which has fallen out of favour recently because congestion is not always present (particularly if the patient is managed well) and biventricular failure is the preferred phrase.

LEFT VENTRICULAR SYSTOLIC DYSFUNCTION (LVSD)

At present the term 'left ventricular systolic dysfunction' is often used, incorrectly, as a surrogate for heart failure. It describes the left ventricle not contracting properly and could be considered a sub-division of left ventricular failure. It is the most common form of heart failure in the developed world because it is the usual pattern of heart failure after myocardial infarction. It is characterised on echocardiogram by non-contracting (akinetic) or poorly contracting (hypokinetic) areas of the left ventricular wall.

The reason why left ventricular systolic dysfunction is widely used as a definition of heart failure is because it is easy to detect on echocardiogram. This makes it a simple marker to look for in trials or as part of screening programmes or registers. For these reasons, confirming left ventricular systolic dysfunction on echocardiogram is used as one of the quality markers in the GMS contract, the system by which GPs are rewarded for meeting health targets in the UK. The problem with using left ventricular systolic function in this way is that it ignores heart failure with preserved left ventricular function – a significant group that includes around 40% of the whole heart failure population.

DIASTOLIC HEART FAILURE

Until relatively recently diastolic heart failure was a controversial definition. This was because it is difficult to find objective evidence for diastolic dysfunction with

the diagnostic tests we have. Diastolic dysfunction occurs when the heart does not relax properly after systole. This may be caused by scarring, hypertension or the fibrotic changes of old age. It is thought that diastolic heart failure may account for the clinically observed fact that some patients seem to have all the signs and symptoms of heart failure but have preserved left ventricular systolic function on echocardiogram (Andrew 2003). It is estimated that 40% of patients with heart failure may have diastolic dysfunction. In the CHARMES study diastolic dysfunction was found on the echocardiogram of two thirds of patients hospitalised with symptoms of heart failure but who had preserved systolic function (Persson et al. 2007).

LEFT VENTRICULAR HYPERTROPHY (LVH)

Although left ventricular hypertrophy is not strictly speaking a term that describes heart failure it is worth considering because of its close physiological links. It describes an increase in the muscle mass of the left ventricle. This is usually caused as a response to either myocardial infarction or chronic hypertension. As a result of these causes, the discovery of left ventricular hypertrophy on an electrocardiogram or echocardiogram is an important finding. Even if no overt heart failure is found the patient is at high risk of developing heart failure in the future and should be treated and followed up closely.

GRADING OF HEART FAILURE

We have seen that there are many definitions available for heart failure. There are also several ways that heart failure can be graded. As well as the structural and descriptive classification discussed earlier it is possible to attempt to grade severity by objective measurements.

One way to do this is to calculate the left ventricular ejection fraction (LVEF) – the percentage of blood that leaves the left ventricle into circulation. As the usual stroke volume is around 70 ml and the typical left ventricular end-diastolic volume 120 ml, the ejection fraction is 70 divided by 120, which is 0.6 or 60%. The ejection fraction is a way of quantifying left ventricular systolic dysfunction. The rationale for being interested in the ejection fraction is that the worse the ejection fraction the more severe the potential heart failure. This has some validity, although it needs to be stressed that individual patients with a poor ejection fraction may be asymptomatic, or a particular patient with a good ejection fraction may have severe symptoms. The degree of reduction of left ventricular ejection fraction is often measured as part of the demographics and inclusion/exclusion criteria of clinical trials in heart failure.

Box 1.5 Left Ventricle Ejection Fraction and Systolic Dysfunction

LVEF %	Systolic Dysfunction
60 %	Normal
45–60 %	Mild left ventricular dysfunction
35–45 %	Moderate left ventricular dysfunction
25–35 %	Severe left ventricular dysfunction
< 25 %	Extremely severe left ventricular dysfunction

A widely used method for classifying heart failure is the New York Heart Association (NYHA) grading. This has been in use since 1928 and provides a four-stage classification based on the patient's symptoms (Subramanian et al. 2005). It has the advantages of being both easy to remember and a simple practical method for testing whether someone has improved or worsened. It is not, however, infallible as some observer bias is inevitable and it relies on patient self-reported information. It is also limited in that it is difficult to assess the NYHA grade in patients with respiratory conditions.

Box 1.6 New York Heart Association (NYHA) Classification

- Class I: Asymptomatic
- Class II: Symptoms on maximal exertion
- Class III: Symptoms on minimal exertion
- Class IV: Symptoms at rest

In 1998 the American Heart Association and the American College of Cardiology introduced a classification scheme to stage heart failure. This is based on the successful staging system used in cancer care. The purpose is to recognise that heart failure is a continuum and to refocus attention on patients who are at risk of heart failure or in the early stages, so that they receive treatment long before they become acutely unwell.

Box 1.7 AHA/ACC Heart Failure Staging Classification

- A No clinical, structural or functional signs of heart failure but at high risk of developing heart failure due to co-morbidities (such as hypertension, ischaemic heart disease, alcoholism, rheumatic fever, etc.)

Box 1.7 (Continued)

- B No clinical signs or symptoms of heart failure but evidence of structural heart disease (such as left ventricular dilation or hypertrophy, previous myocardial infarction or valve disease)
- C Symptoms of heart failure and underlying structural heart disease
- D Symptoms of heart failure at rest, despite treatment optimisation, with advanced structural heart disease

(Hunt et al. 2001)