Chapter 25

Structural heart disease

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Learning Objectives

After reading this chapter and completing the learning activities, the reader will be able to:

- Describe cardiac conditions classed as acquired and hereditary structural heart disease
- Discuss the relationship between rheumatic heart disease and valvular disease
- Identify the most common forms of cardiomyopathy and their causes
- Discuss the role of the nurse in a multidisciplinary team caring for patients with structural heart disease
- Identify sources of information for patients with strutural heart disease and their families.

Key Words

Valvular disease, valve stenosis, valve regurgitation, cardiomyopathy, rheumatic heart disease

Overview

Structural heart disease is the term used to describe conditions that affect the valves, muscles, or walls of the heart. Structural heart disease may be present from birth (congenital), but they often can develop with ageing or result from other diseases (acquired). This chapter provides an overview of common structural heart diseases. Congenital heart disease is discussed in detail Chapter 24.

Valvular heart disease

The most common form of structural heart disease is valvular disease. The prevalence of VHD is estimated to be around 2.5% in high income countries (Lung and Vahanian, 2014). With the decline of rheumatic heart disease (RHD), degenerative aetiologies have become the predominant cause of VHD in high income countries resulting in a marked increase in diagnosis in patients over the age of 65 (Lung and Vahanian, 2014).

Valvular heart disease (VHD) is caused by either damage or defect in one of the four heart valves: aortic; mitral; tricuspid; or pulmonary. VHD may present as valvular stenosis, regurgitation, or a combination of both. A stenotic valve is one where the valve acts as an obstruction to blood leaving the heart chamber, whilst regurgitation, or valve insufficiency, refers to leakage of blood through the valve as it closes after each heartbeat. Aortic stenosis and mitral regurgitation are the most common diagnoses, continuing to account for about 3 in 4 cases of VHD (Lung and Vahanian, 2011, Bravo-Jaimes et al., 2018). Valvular abnormalities may be congenital or acquired. This section focusses on acquired valve disease. For in-depth information on current management strategies for valvular disease, it is recommended that you consult the appropriate national/international guidelines used in your country.

Key Point

Endocarditis prophylaxis with antibiotics should be considered for high-risk procedures in patients with prosthetic valves, including transcatheter valves, or with repairs using prosthetic material, and for those with previous episodes of infective endocarditis (IE) (Baumgartner et al. 2017). For more information on endocarditis prophylaxis, see Chapter 24.

Suggested resources: Guidelines

- Otto, C.M., *et al*. (2020). 2020 ACC/AHA Guideline for the Management of Patients with Valvular Heart Disease: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. https://doi.org/10.1161/CIR.0000000000000923
- Baumgartner, H. *et al.* (2017). 2017 ESC/EACTS guidelines for the management of valvular heart disease. *European Heart Journal*, *38*(36), 2739-2791. https://doi:10.1093/eurheartj/ehx391

Box 2?.1 Rheumatic valvular heart disease

Acute rheumatic fever (ARF) and its sequel, rheumatic heart disease (RHD) are under-recognised conditions that are associated with significant morbidity and mortality in developing countries and in some sub-populations in developed countries (Watkins et al 2017). Infections such as strep throat, scarlet fever, or skin sores, pyoderma, impetigo that are associated with Group A β- haemolytic streptococcus (GAS) can lead to ARF. In a

susceptible host GAS may trigger an autoimmune response that results in a generalised inflammatory illness (ARF). Generalised inflammatory symptoms that involve the skin and joints may resolve, but if the heart has been affected (acute carditis), damage to the heart valves may remain once the acute carditis has resolved, leading to chronic RHD. Recurrent GAS infections and episodes of ARF can further damage the heart valves, making RHD progressively worse over time (He et al. 2016). Prevention of RHD at the time of a first attack of acute rheumatic fever is the best strategy. In the case of infection, antibiotic treatment of group A Streptococcus sore throat is key in primary prevention. Secondary national or international guidelines for management of RHD (see suggested resource guidelines above) provide advice on secondary prophylaxis regimes, antibiotic choice, dose, frequency, and duration of secondary prophylaxis.

Key Point Rates of RHD among Indigenous (Aboriginal and Torres Strait Islander) Australians, many of whom live in poverty in overcrowded conditions, are among the highest rates reported worldwide (He et al. 2016).

Aortic valve disease

The aortic valve lies between the left ventricle and the aorta. During the cardiac cycle, the left ventricle contracts and the blood is expelled through the aortic valve into the aorta and around the systemic circulation.

Aortic valve stenosis

Aortic valve stenosis (AVS) is the most common primary valve disease that leads to surgery or catheter intervention in Europe and North America (Baumgartner et al. 2017). In patients with aortic stenosis, the afterload (the amount of resistance the heart must overcome to open the aortic valve and push the blood volume out into the systemic circulation) steadily increases as the aortic valve narrows. As a result, the left ventricle is required to generate an increasingly high systolic pressure to pump the blood through the valve. To compensate for this, the left ventricular (LV) muscle wall thickens. This is known as concentric hypertrophy (Carabello & Paulus, 2009). The thickened myocardium enables the LV systolic contraction to strengthen, thus initially maintaining an adequate stroke volume (SV) and cardiac output (CO). However, over time, compensatory mechanisms start to fail, heart failure (HF) develops (see Chapter 22 for the pathophysiology of HF). The hypertrophic myocardium may

also compress the coronary arteries, thereby reducing coronary blood flow and potentially causing ischaemia.

Aortic valve replacement (AVR) is the only 'curative' intervention for AVS but the decision to offer intervention must be carefully considered: to intervene too early may unnecessarily expose patients to the risks of valve replacement; to intervene too late may cause irreversible cardiac damage that is associated with an increased risk of HF and death (Bing & Dweck 2020). AVR may be performed using transcatheter aortic valve implantation (TAVI) or surgical AVR according to the patient risk profile and availability of the intervention. A recent metanalysis of trials comparing surgical AVR with TAVI suggests that in patients with symptomatic, severe AS, TAVI was associated with a reduction in all-cause mortality and stroke up to 2 years compared surgical AVR (Siontis et al. 2019). However, long-term durability data of TAVI beyond 5 years are still limited .

Aortic regurgitation

Aortic regurgitation (AR) can be caused by primary disease of the aortic valve cusps and/or abnormalities of the aortic root and ascending aortic geometry. In Western countries, AR is more commonly due to degenerative causes. Acute AR may occur secondary to infective and rheumatic endocarditis, and less commonly, aortic dissection, blunt chest trauma or iatrogenic causes following transcatheter procedure (Otto et al. 2020). Asymptomatic patients with severe aortic regurgitation require careful follow-up of symptomatic status and LV size and function (Baumgartner et al. 2017). Valve surgery is indicated for symptomatic patients (spontaneous or on exercise testing) and/or LVEF <50% and/or end-systolic diameter>50mm. Medical therapy can provide some symptomatic improvement in individuals with chronic severe AR in whom surgery is not feasible (Otto et al 2020). In patients with acute severe aortic regurgitation from aortic dissection or infective endocarditis, medical therapy to reduce LV afterload may be required, especially if hypotension, pulmonary oedema, or evidence of low flow is present (Otto et al. 2020).

Key Point Intra-aortic balloon counterpulsation (IABP) is contraindicated in patients with acute severe AR (Otto et al 2020).

Mitral valve disease

The mitral valve sits between the left atrium and left ventricle. In a normal cardiac cycle, the mitral valve opens as the atrium contracts and blood flows through into the ventricle. The valve

then closes during systole as the left ventricle contracts. The mitral valve is functionally and anatomically complex and consists of several components: the annulus, the leaflets and the sub-valvular apparatus including the chordae tendineae, the papillary muscles and the ventricular wall (Del Forno et al. 2020).

Mitral regurgitation

Mitral regurgitation (MR) is the second-most frequent indication for valve surgery in Europe (Baumgartner et al. 2017). As with aortic valve disease, degenerative mitral valve disease currently accounts for the majority (60%) of cases, with rheumatic valve disease accounting for around 12% of cases. Ischaemic mitral valve disease currently accounts for around 25% of cases, with the number of cases increasing annually (Izquierdo-Gomez et al., 2018).

It is essential to distinguish primary from secondary MR as the two diseases have different aetiologies, pathophysiology, natural history, and thus different indications and responses to medical, transcatheter, and surgical therapies. Primary MR is caused by inherent lesions that impair the function of the mitral apparatus, including the leaflets, chordae tendineae, papillary muscles, or annulus, and is the most common cause of primary MR is mitral valve prolapse. As primary MR is due to disease of the valve itself, treating the valve can be curative.

In secondary MR, the mitral valve is morphologically normal, but disease of the left ventricle has led to abnormal anatomy and function that causes displacement of one or both papillary muscles, leaflet tethering, inadequate closure of the valve, and often annular dilatation. Secondary MR may result from ischemic or nonischemic ventricular disease including ischaemic cardiomyopathy, idiopathic dilated cardiomyopathy, atrial functional MR or hypertrophic cardiomyopathy. Atrial fibrillation with left atrial enlargement may be the underlying mechanism of secondary MR when LV structure and function are normal (Del Forno et al. 2020). In secondary MR, the mitral valve itself is not the origin of the disease, therefore therapy directed only at MR may reduce regurgitation but cannot cure the basic underlying pathology.

Key Point	In chronic MR, the left ventricle adapts for some time, and symptoms may be delayed for years (Lung, 2003). However, in acute MR, the left ventricle is unable to adequately compensate, and the patient is likely to present with severe symptoms. This is reflected in increased mortality and morbidity rates (Nishino et al., 2016.)

Mitral stenosis

Mitral stenosis (MS) is characterised by obstruction of blood flow across the mitral valve from the left atrium to the left ventricle (Mitral stenosis has a variable presentation. RHD has been a frequent cause of mitral stenosis (MS) worldwide, though the incidence is low in high-income countries, and slowly declining elsewhere. In regions with a high prevalence of RHD, patients usually present at a younger age (teen years to 30 years) (Otto et al. 2020). Another infrequent form of MS may result from congenital heart disease. Non-rheumatic degenerative calcific mitral valve disease is encountered mainly in elderly patients (Baumgartner et al. 2017). Calcific MS results from calcification of the mitral valve annulus that extends top the base of the leaflets resulting in narrowing of the annulus and rigidity of the leaflets (Gaasch 2020).

One of the most common complications of MS is atrial fibrillation (AF) (Lung et al., 2018). AF is associated with an increased risk of morbidity, in particular stroke. Increased pressure in the left atrium leads to marked structural and electrical remodeling, which in turn predisposes the atrium to tachyarrhythmias. AF also reduces the haemodynamic tolerance of mitral stenosis, through the loss of the atrial kick. Oral anticoagulant therapy is essential when AF complicates mitral stenosis, regardless of its severity.

Key Point	Medical management of atrial fibrillation (AF) does not differ significantly from routine AF guidelines, but as AF often has exaggerated haemodynamic effects in MS, prompt therapy is often necessary (Gaasch 2020).

Periodic monitoring is recommended in asymptomatic patients with MS to assess for disease progression and development of indications for intervention (Gaasch 2020). Diuretics, beta-blockers, digoxin or heart rate–regulating calcium channel blockers can transiently improve symptoms. The timing and type of intervention is decided based on clinical characteristics, valve anatomy and local expertise. Percutaneous mitral commissurotomy should be considered as an initial treatment for selected patients with mild to moderate disease who have otherwise favourable clinical characteristics and surgery, usually for valve replacement, is indicated in the other patients (Baumgartner et al. 2017).

Tricuspid valve disease

The tricuspid valve sits between the right atrium and right ventricle. It opens as the atria pump, then closes as the ventricles contract, to ensure that blood flows in a forward direction. This enables blood to be pumped from the right side of the heart to the lungs for oxygenation.

Tricuspid regurgitation

The more common form of tricuspid disease is tricuspid regurgitation (TR). This occurs when the valve cannot close adequately, resulting in the backward leakage of blood into the right atrium with each heart contraction. Causes of tricuspid regurgitation include HF (which can be a cause or a consequence of TR), cardiomyopathies, ventricular dilation secondary to high blood pressure in the pulmonary circulation (pulmonary hypertension), and less commonly, trauma, infective endocarditis, RHD, carcinoid syndrome and congenital heart defects (Harris et al. 2017). For further information on congenital heart defects, refer to Chapter 24.

Tricuspid stenosis

In tricuspid stenosis (TS), the valve becomes stiff and narrowed, preventing the forward flow of blood from the atrium to the ventricle (Harris et al. 2017). TS most commonly results from RHD, which causes the leaflets of the valve to become thick, hardened and less able to open widely, thus restricting forward blood flow.

Key Point	Infective endocarditis on the right side of the heart is relatively rare (Shrestha et al., 2015). But around 90 - 95% of cases involve the tricuspid valve (Murdoch et al., 2009). Many of these infections result from IV drug use, accounting for approximately 30- 40% of tricuspid valve endocarditis (Edmond et al., 2001; Hussain et al., 2017).

Patients commonly present only when the valve disease is severe. Symptoms reflect those of right sided HF, and include fatigue, shortness of breath, raised central venous pressure and potentially an enlarged pulsating liver. Management of tricuspid valve disease will depend on the severity of the disease and symptoms as well as the underlying cause. Treatment of the underlying HF with medication can relieve symptoms. Valve repair or replacement may be an option depending on co-morbidities (Harris et al., 2017).

Pulmonary Valve Disease

The pulmonary valve sits between the right ventricle and the pulmonary artery. Abnormalities of the pulmonary valve may lead to an obstruction to the right ventricle outflow tract.

Pulmonary valve stenosis

Pulmonary valve stenosis (PVS) is predominantly associated with congenital cardiac disease, but in rare instances, acquired disorders such as carcinoid or rheumatic fever affect the pulmonary valve (Fathallah et al. 2017). PVS results in overload of the right ventricle, which in turn leads to compensatory right ventricular hypertrophy. Increased muscle mass is a compensatory mechanism to assist the right ventricle to maintain a normal CO. Over time,

progressive right ventricular hypertrophy can give rise to right ventricular dysfunction. Treatment options for PVS include surgical valvotomy, balloon pulmonary valvuloplasty (BPV) and pulmonary valve replacement. Balloon pulmonary valvuloplasty (BPV) is the procedure of choice for PVS as it is less invasive and has a very high success Long-term complications following BPV are progressive PR and restenosis
(Fathalla et al. 2017).

Pulmonary valve regurgitation

Pulmonary regurgitation (PR) is rarely seen as a primary pathology. Primary causes of PR have some overlap with causes of PS, but PR is most often the result of prior intervention on the pulmonary valve (Ruckdeschel & Kim, 2019). Iatrogenic PR may occur after surgical valvotomy, percutaneous valvuloplasty or Tetralogy of Fallot-related surgery (Fathalla et al. 2017).

Key Point	Valvular issues resulting in stenosis and/or regurgitation are established long-term consequences of mediastinal radiation (Ky et al. 2020).

Cardiomyopathy

Cardiomyopathies are disorders of the cardiac muscle that are characterised by mechanical and/or electrical dysfunction that result in dilated, hypertrophic or restrictive pathophysiology (Schultheiss et al. 2019). Cardiomyopathy can be classed as being primary or secondary. Primary cardiomyopathy does not result from another disease or condition that leads to a weakened heart muscle. In some cases, the cardiomyopathy is inherited and may be passed down to other family members. Secondary cardiomyopathy may result from a medical condition such as hypertension, valve disease, congenital heart disease, coronary artery disease, illicit drug use, excessive alcohol use, chemotherapy and other medications. The goal of therapy for patients with secondary cardiomyopathy is to identify and correct the medical condition(s) that are responsible for the condition.

Initially, patients with a cardiomyopathy may experience few or no symptoms. However, longer-term, patients are likely to experience symptoms associated with HF. Many develop arrhythmias, and are subsequently at higher risk of sudden cardiac death (Bagnall, Weintraub et al. 2016).

Primary cardiomyopathies are classified into several general categories depending upon their respective disorders of structure and function. The most common of which are dilated

cardiomyopathy (DCM), hypertrophic cardiomyopathy (HCM) and arrhythmogenic right ventricular dysplasia cardiomyopathy (ARVD/C).

Key Point Cardiomyopathy is a group of conditions with varying pathophysiology and underlying causes. While treatment for symptoms of cardiomyopathy-related HF is similar, it is important to differentiate the aetiology to ensure effective management of the condition.

Dilated Cardiomyopathy

Dilated cardiomyopathy (DCM) is one of the most common causes of HF and the most common indication for heart transplantation worldwide (Weintraub et al. 2017). DCM is characterised by LV or biventricular dilation and impaired contraction that is not explained by abnormal loading conditions (such as hypertension and valvular heart disease) or coronary artery disease. There are several causes of DCM. Around 50% of cases are idiopathic and around half of these patients will have familial disease. DCM can be a component of an inherited disorder, such as neuromuscular diseases (muscular dystrophies and myotonic dystrophy), hereditary haemochromatosis, and hereditary sideroblastic anaemias and thalassaemias. There are currently no clinical or histologic criteria, other than family history that distinguish familial from nonfamilial disease (Weigner & Morgan 2020). Secondary causes include myocarditis, ischaemic heart disease, infiltrative disease (amyloidosis, sarcoidosis, and haemochromatosis), peripartum cardiomyopathy, autoimmune conditions, endocrine disorders (thyroid dysfunction, pheochromocytoma), hypertension, HIV infection, connective tissue disease, chemotherapy agents, and illicit drug and heavy alcohol use (Japp et al., 2016; Schultheiss et al. 2019; Weigner & Morgan 2020). Standard approaches to prevent or treat HF are the first-line treatment for patients with DCM (see Chapter 23), including cardiac resynchronisation therapy and implantable cardioverter–defibrillators (see Chapter 12) to prevent life-threatening arrhythmias if required (Schultheiss et al. 2019). Identification of the probable cause of DCM helps tailor specific therapies to improve prognosis (Schultheiss et al. 2019).

Genetic testing

Genetic testing in patients with DCM facilitates screening and all first-degree relatives of patients with familial DCM should undergo clinical screening. Pre-test counselling needs to be provided to all patients with DCM, ideally by a person with knowledge of DCM

genetics, such as a genetic counsellor. Pre-test counselling helps to facilitate patient understanding of the rationale for testing, increase empowerment, and improve communication and dynamics within families. Patients must also understand issues relating to the privacy of their genetic testing results and how it may impact on aspects of their lives in areas such as employment and insurability for themselves and their family members (Rosenbaum et al. 2020).

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is the most common heritable cardiomyopathy though the underlying genetic cause of disease is only found in 34% of patients (Bos et al. 2014). The LV becomes hypertrophic (thickened), non-dilated in HCM and left ventricular (LV) function is preserved (Marian & Braunwald 2017). Patient may be asymptomatic or present with HF or sudden cardiac death (SCD). HCM can be classified as obstructive or non-obstructive depending on whether the heart anatomy causes left ventricular outflow obstruction (LVOT) (Geske et al. 2018). Symptoms relate to the type of structural or functional abnormalities and include fatigue, dyspnoea, chest pain, palpitations, and presyncope or syncope (Maron 2020). Therapy is aimed at reduction of LVOT and includes lifestyle modifications, pharmacotherapies, and septal reduction therapies. Most patients experience little or no disability, and life expectancy is normal. However, a small subset of patients with HCM will experience SCD (around 1% annually), with risk stratification a significant clinical challenge (Geske et al., 2018) and concordance in international guidelines is lacking. High-risk patients may be treated effectively with the implantable cardioverter-defibrillator. Genetic testing in HCM largely centers on family screening; if a causative genetic mutation is identified, testing for this mutation is the preferred method of family screening (Geske et al. 2018).

Restrictive cardiomyopathy

Restrictive cardiomyopathy is rare condition. Restrictive cardiomyopathy may occur secondary to causes such as amyloidosis, haemochromatosis, and sarcoidosis, but the cause is often idiopathic. Characteristics of restrictive cardiomyopathy that distinguish it from other forms of cardiomyopathy include a non-dilated ventricle with typically normal wall thicknesses, rigid ventricular walls that result in severe diastolic dysfunction and dilated atria, and generally normal LV systolic function (Ammash & Tajik, 2020). Some people with this condition have no symptoms or minor symptoms, but others may develop symptoms of HF and arrhythmia.

There is no specific therapy for idiopathic restrictive cardiomyopathy. Management includes lifestyle modifications and standard approaches to HF management. Treatment of the disease may be beneficial in patients with secondary restrictive cardiomyopathy (Ammash & Tajik, 2020).

Key Point Radiation therapy to myocardial tissues can result in a restrictive cardiomyopathy (Ky et al. 2020).

Arrhythmogenic right ventricular dysplasia cardiomyopathy

ARVD/C is a rare inherited form of cardiomyopathy in which the right ventricular heart muscle is gradually replaced by fat or fibrous tissue. As a result, the right ventricle becomes dilated, with reduced contractility. ARVD is characterised by ventricular tachyarrhythmia, predominant right ventricular dysfunction, and sudden cardiac death (Wang et al. 2017). The estimated prevalence of ARVD ranges from 1 in 5000 to 1 in 2000 in some European countries (Coelho et al., 2019). Patients typically present in the 2nd to 4th decade of life (Wang et al. 2017), usually with palpitations (30–60%), lightheadedness (20%), and syncope (10–30%) linked to the presence of non-sustained or sustained ventricular arrhythmias. Up to 19% of ARVD/C patients present with cardiac arrest. Atrial arrhythmias and atrial fibrillation have also been associated with ARVD/C (Wang et al 2017). ARVD is one of the leading causes of arrhythmic cardiac arrest in young people and in particular, athletes (Basso et al., 2009). Whilst over 50% of diagnoses have a genetic origin, ARVD may also occur due to non-genetic causes such as myocarditis, or congenital abnormalities affecting the right ventricle.

The clinical approach to managing patients with ARVD/C patients consists of establishing an accurate diagnosis; risk stratification for sudden death; prevention of sustained ventricular arrhythmia; preventing the development of HF; cardiac transplant; and screening and follow-up of family members. ICD implantation with anti-tachycardia pacing (ATP) may be recommended following a discussion about the risk-benefits of having/not having and ICD. The use of anti-arrhythmic drugs rarely eliminates the risk of sudden death and is mainly to reduce the arrhythmia burden (Wang et al. 2017). See Chapter xx for further information on AVRD.

Key Point Men have an earlier onset of AVRD and once diagnosed, have worse outcomes than women. It is thought that this may be due sex-related difference in hormone profiles and exercise participation (James et al., 2013; Wang et al. 2018).

Learning activity 24.1

When patients receive a serious diagnosis such as arrhythmogenic right ventricular dysplasia cardiomyopathy, they generally go to the internet for information. Identify at least three reliable and evidence-based sources of information that are suitable for patients with conditions discussed in this chapter.

Other forms of cardiomyopathy

Some types of cardiomyopathy do not fit well into the general classifications. These are discussed briefly below.

Takotsubo syndrome

Takotsubo syndrome (TTS), previously known as 'takotsubo cardiomyopathy', 'stress cardiomyopathy' or apical ballooning syndrome' is a condition that is usually associated with an emotional or physical stressor that leads to transient LV wall motion abnormalities and reduced EF that may lead to HF (see Chapter xx).

Peripartum cardiomyopathy

Peripartum cardiomyopathy is a rare form of cardiomyopathy with systolic dysfunction that typically presents in late pregnancy and up to six months postpartum. The cause of PPCM is unknown and the diagnosis of PPCM is one of exclusion of other causes of HF. PPCM incidence is highest in women of African descent and other associated factors include age >25 years, pregnancy associated hypertensive disorders, and anaemia (Rodriguez Ziccardi 2020). The LV ejection fraction (EF) is almost always below 45% and the left ventricle may or may not be dilated. PPCM is associated with HF, an increased risk of atrial and ventricular arrhythmias, thromboembolism, and sudden cardiac death. The initial medical management of PPCM is like that for other causes of HF, with the special consideration of how the pregnancy will be affected.

Therapeutic considerations may include arrhythmia management, anticoagulation therapy, inotropes (with close monitoring and weaning off as soon as practical), mechanical support, and possible transplant (Rodriguez Ziccardi 2020). More than half of affected women recover systolic function, although some are left with a chronic cardiomyopathy, and a minority require mechanical support or cardiac transplantation (Honigberg & Givertz 2019). Women with a minimal decrease in EF tend to have a good prognosis, whereas those with a poor EF and those requiring mechanical support have a higher risk of death (Rodriguez Ziccardi

2020). Information, education, and support is critical for patients with PPCM. As management of PPCM involves input from various specialties (obstetrics, midwifery, cardiology, heart failure teams; social work, community health and possibly mental health), coordinated multidisciplinary support is essential for these patients. Many will face significant physical and psychological challenges in caring for a newborn whilst they are ill.

Key Point A recent study has suggested that around one third of pregnancy-associated cardiomyopathy patients had TTS. Peripartum takosubo syndrome should be considered as a differential diagnosis of HF during peripartum (Kim et al. 2020).

Chemotherapy/radiotherapy-induced cardiomyopathy

Cancer therapy may result in damage to multiple cardiac structures as a late complication that may not be manifest for years after treatment has completed. The outcomes of cancer survivors who develop cardiomyopathy (whether due to chemotherapy or radiation therapy) are generally poor. Anthracycline analogues, particularly doxorubicin (Adriamycin), are the chemotherapeutic agents most associated with cardiotoxicity, but there are several others (Ky et al. 2020). Historically, it has been thought that cardiotoxicity related to anthracyclines is irreversible, but more recent evidence demonstrates that early identification of cardiotoxicity and prompt therapy for HF can lead to substantial improvement in LVEF. Early detection of cardiac dysfunction and provision of cardiac-specific therapy is imperative to prevent progression of cardiac dysfunction (Ky et al. 2020).

Key Point It has been suggested that chemotherapy accounts for 1–2% of TTS triggers (Storey & Sharkey 2019). Fluorouracil (5FU) is the most reported, though there are many others (Coen et al. 2017). The onset of TTS relative to chemotherapy initiation is quite variable and may occur at any time from the initial administration to several weeks beyond initiation. TTS in the setting of treatment for malignancy is associated with substantial mortality (Storey & Sharkey 2019).

The multidisciplinary team caring for patients with acquired structural heart disease

A multidisciplinary team approach inclusive of cardiologists, surgeons, specialist nurses, physiologists or clinical scientists is recommended for all types of valve disease and infective

endocarditis (Chambers et al 2017). Roles of the nurse may include providing information and education for the patient about their condition, monitoring of the patient for symptoms, physical examination, review of medication, lifestyle advice about smoking cessation, weight control and dental surveillance and communicate the results of follow-up with the general practitioner and referring physician. Nurses often coordinate outpatient clinics, manage appointments, maintain audit databases, follow up laboratory results, and run the telephone and email helplines. Multidisciplinary valve clinics have been found to be patient-focused and cost-effective (Chambers et al. 2020).

Patients with cardiomyopathy complicated by HF are also best managed by a multidisciplinary team with health care professionals that specialise is various aspects of care, including primary physicians (general practitioners), HF nurse specialist, cardiologist, dietician, pharmacist, psychologist, physical and occupational therapists, exercise physiologist, and social worker. Additionally, patients with familial DCM and their families may needed additional support from geneticists and councillors.

Conclusion

Structural heart disease encompasses a broad range of congenital, hereditary, and acquired conditions with a wide variation in morbidity and mortality. Structural heart disease can present across the lifespan and occur as a primary condition or secondary to other disease processes, environmental and lifestyle factors. Heart failure is a common feature of many structural heart diseases and early identification and intervention is needed to halt the progression of disease and maximise cardiac function to improve survival and quality of life.

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