A PATIENT GUIDE TO HEART VALVE DISEASE

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Edited by Patricia Khan

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A PATIENT GUIDE TO HEART VALVE DISEASE AND HEART VALVE DISEASE





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Edited by Patricia Khan

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John Chambers is Professor of Clinical Cardiology and Consultant Cardiologist at Guy's and St Thomas's Hospital, London. A founding member and inaugural President of the British Heart Valve Society, he is the pioneer of specialist heart valve clinics in the UK.

This Patient Guide is a summary of a lifetime's work and a career-long interest in heart valve disease. It explains how normal heart valves function, what causes valve disease, what symptoms it can produce and how the disease is detected and treated.

It covers every type of valve disease, what questions to ask your surgeon, and how to prepare for surgery.

"My goal for this Guide is to offer a reliable go-to resource other than the internet for any non-medical person keen to know more about heart valve disease.

You may be a patient, a relative, a carer, or a patient yet to be diagnosed. Whatever the reason for your interest, you want reliable information delivered in plain English so that you don't need to reach for reference books.

Heart valve disease is increasingly common in the U.K. and in Europe. This Guide should help you to understand the condition, and give you the information you need to make decisions about your care, or a loved-one's.

Methods of treatment are advancing rapidly, and opportunities for patients to take part in decisions about their care are often welcomed and encouraged by medical teams. I sincerely hope this Guide will help you in that process."

Chapter One How do Heart Valves Work?

HOW HEART VALVES WORK

The heart is a pump with two parallel circuits, right and left. Heart valves open to allow blood to flow in the correct direction, and close to stop it travelling backwards.



Blood from the body enters the right atrium, is passed to the right ventricle, then on to the lungs where it picks up oxygen.

Oxygenated blood from the lungs enters the left atrium, is passed to the left ventricle, then out via the aorta to the head and all the organs of the body. When the left and right ventricles fill with blood, there is a momentary pause before the ventricles contract or close to eject the blood.



The mitral valve is the inlet valve on the left circuit. This opens to let blood fill the left ventricle, then closes to stop blood flowing backwards into the left atrium. If the mitral valve isn't working properly and blood does flow backwards, this is called **reflux**, or **mitral regurgitation**.

As the mitral valve closes, the aortic valve opens to let blood flow from the left ventricle into the aorta, and onwards around the body.







The tricuspid valve is the inlet valve on the right circuit. This opens to let blood flow from the right atrium to right ventricle, then closes to stop blood flowing backwards into the right atrium. If the tricuspid valve isn't working properly and blood does flow backwards, this is called **tricuspid regurgitation**.

As the tricuspid valve closes, the pulmonary valve opens to let blood flow from the right ventricle into the pulmonary artery and on to the lungs.





Mitral valve

The mitral valve has two leaflets, front and back. These leaflets overlap when closing by 5 - 10 mm. Muscles attached to the middle segments of the left ventricle support the leaflets, and prevent them from flailing backwards into the left atrium. If this flailing or slipping does occur, it is called prolapse.

It is normal for a small amount of blood to leak backwards as the mitral valve closes. **Abnormal mitral regurgitation** can occur when any part of the mitral valve isn't working properly.

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Aortic valve

The aortic valve consists of three cusps mounted at the base of the onion-like portion of the aorta. These cusps open and close in response to pressure from blood flow.

Aortic regurgitation occurs as a result of abnormal widening of the aorta, or abnormalities of the aortic valve, or a mixture of both.



Tricuspid valve

The tricuspid valve consists of three leaflets. This valve is thinner/more delicate/less robust than the mitral valve. However, normal tricuspid regurgitation is more common than normal mitral regurgitation.

Abnormal tricuspid regurgitation occurs because of excessive widening at the base of the valve, or of the right ventricle, or because of abnormalities of the leaflets themselves.



Pulmonary valve

The normal pulmonary valve consists of three cusps. These cusps open and close in response to pressure from blood flow.

Chapter Two What is Heart Valve Disease?

VALVE DISEASE EXPLAINED

- Firstly, it's not a 'disease' like scarlet fever. It's a dis-function. What happens is that the valves of the heart get thicker, or weaker, and can't pump or push blood as well as they should
- The aortic and mitral valves are most often affected
- Heart valve disease can occur naturally as people grow older
- There is no exact point at which normal thickening or weakening becomes 'valve disease,' but there are generally accepted thresholds
- It can be put right with a mechanical solution like replacing a washer or a pipe.

If a valve in your heart becomes thickened or weakened it will fail to open properly, or it will leak.



Failing to open is called **stenosis**

Leaking is called regurgitation

Mild regurgitation is very common

Severe cases need surgery

In the over-65s, the most common valve diseases are aortic stenosis and mitral regurgitation. Approximately one in three people have thickening of the aortic valve.

In the over-80s, one in five people have mitral valve thickening.

Stenosis or **regurgitation** or sometimes a mixture of the two, is regarded as **'significant valve disease'**. This needs regular monitoring in a specialist clinic.

Aortic regurgitation is less common.

Mitral stenosis and diseases of the tricuspid and pulmonary valves are even more uncommon.

What causes valve disease?

The causes depend largely on where you live in the world, and whether you have access to good housing, drainage and medical care.

Wherever you live, there are a number of common causes; bicuspid aortic valve, mitral prolapse and congenital anomalies. These are discussed in more detail in other chapters.

Worldwide, the most common cause of valve disease is rheumatic fever, contracted between the ages of 5-15 by people living mostly

in poverty-stricken areas of South America, India and Africa.

Rheumatic fever usually follows a very bad streptococcal throat infection. Antibodies rush to the body's defence, including to the tissues of the heart. Multiple episodes are usually needed to



cause permanent damage to the heart valves, which would become scarred and thickened over years.

The worst affected country is Ethiopia, where 20% of people with rheumatic disease die before the age of 5, and 80% before 25.

Even in endemic areas, the prevalence varies widely. In a study from Africa the prevalence was 2.7% in school children in a lower-income area of Cape Town compared with only 1.25% in a higher-income area. In the same study the average prevalence was 2% in South Africa compared to 3% in Ethiopia which has a poorer more rural population. Since the second half of the 20th century it has become uncommon in industrialised countries as a result in improvement in housing and health care. At the same time these advances have led to an increase in life-expectancy in industrialised countries. Valve disease is increasingly caused by diseases of old age, calcific aortic stenosis and mitral regurgitation caused by LV dysfunction. There are also diseases caused by medical interventions including radiation, some drugs and infection introduced with indwelling venous cannulae. These diseases are also seen in richer areas of the under industrialised nations.

How is valve disease treated?

Early detection is really important.

It is possible to delay, even prevent, early thickening or leakage of heart valves caused by rheumatic fever, simply by using penicillin.



If heart valve damage becomes severe, surgery is usually the only effective treatment.

In the past, cases tended to be picked up only when the damage to the valves had become severe, or when a murmur was detected by a doctor using a stethoscope.

With the push to establish more specialist valve clinics across the UK (*see chapter 18*), people with valve disease can be followed and monitored more closely.

This means they will be given the appropriate treatment and care to live well until the right time for surgery can be judged correctly.

How common is valve disease?

The frequency of a condition is described in two ways:

Prevalence

This means the burden of disease present at any one time expressed as a proportion of the population e.g. % or per million

Incidence

This means the rate at which new cases are identified expressed as a proportion of the population over a time e.g. per million per year



The British OxVALVE study

Reproduced with permission from d'Arcy JL, Coffey S, Loudon MA, et al. Large-scale community echocardiographic screening reveals a major burden of undiagnosed valvular heart disease inolder people: the OxVALVE Population Cohort Study. *Europ Heart J. doi:10.1093/eurheartj/ehw229*

The British OxVALVE study was aimed at detecting all grades of valve disease in the Oxford area in a target age group. Two thousand five hundred (2,500) people aged 65 and under were invited to attend for an echocardiogram at four GP surgeries. Mild valve disease was found in 44%. Just over 10% had significant valve disease. For those aged 65 and over 19% had significant valve disease.

These figures show that mild 'disease' or dis-function of the heart valves is so common, it is really a part of normal aging. The generic use of the term disease is therefore inappropriate, even alarmist, and should only be applied to cases of severe thickening or leakage.

The Euro Heart Survey

The Euro Heart Survey, a study of 4,910 inpatients or outpatients at hospitals across Europe, showed the frequency of different types of valve disease.



How is valve disease detected?

In the **OxVALVE study**, 4.9% of people were already known to have valve disease, but a further 6.4% had previously undiagnosed moderate or severe valve disease.

GPs using stethoscopes are often the first people to suspect heart valve disease and start the ball rolling towards a diagnosis. We looked at 'open access' (GP requested) echocardiograms performed at Guy's and St Thomas' Hospitals. About half of these tests were to investigate a murmur picked up by a GP, and in these tests 29% of patients were found to have valve disease (mild in 11% and significant in 18%). Almost as many patients without a murmur also had valve disease.

A similar message comes from a large study of 79,043 people with suspected heart failure referred by their GPs to a private company in the UK (Marciniac). The proportion of people with valve disease was substantially higher than with LV damage consistent with heart failure.



The proportion of 79,043 people with suspected heart failure who had valve disease referred by their GP to a private echocardiography company

So, mild valve disease is common, can be normal, and will always be detected if an echocardiogram is performed.

It is also clear that significant valve disease (which needs follow-up in an expert clinic and may potentially require surgery) can also be unsuspected.

- Valve disease is typically thought to cause a murmur
- We know that echocardiography has to be used more widely to detect valve disease (Arden)
- It has to be used in people with significant breathlessness and abnormal rhythms (particularly atrial fibrillation).

This is discussed in more detail in chapter 18.

Frequently Asked Questions

Q: My doctor says I have a murmur. What is this?

A murmur is simply the noise made by blood passing through the heart and is often normal. The pulmonary artery is relatively close to the skin and blood flow may be audible through a stethoscope particularly if you are slim. This is called a 'benign systolic flow murmur'. About 20-30% of all people have a murmur but only about 2-3% have valve disease.

This benign murmur may also be accentuated if the action of the heart is increased for example because you are anxious, have recently exercised or are anaemic or pregnant. A murmur during pregnancy is very common indeed. Usually a benign flow murmur can be differentiated from an abnormal murmur but if there is doubt then an echocardiogram may be necessary.

Q: The echocardiogram report describes mild leakage from some of the valves. Should I be concerned?

Normal heart valves often have slightly irregular edges and it can be normal for these to leak slightly. Mild leakage is usual from the tricuspid and pulmonary valves and may also be normal through the mitral valve.

As we age, minor thickening of the valves develops particularly in the aortic valve. A small degree of leakage of the aortic valve is common especially after about 65 years of age. If there is more than mild leakage you will need to discuss with your doctor whether this is of no consequence or whether further follow-up could be necessary.

Q: Is valve disease infectious?

Disease may sound like an infection but is a term used for any significant change from normal. Valve disease means that the valve has developed abnormally or that it has developed abnormal thickening or become damaged.

It is possible for a leaking or thickened valve to become infected. This is rare. The most common infecting organism is a Streptoccus from the mouth and infection can be avoided by regular dental surveillance. There are also some infectious causes of valve disease in some countries in the world, most commonly rheumatic fever. This is caused by repeated streptococcal throat infections.

You cannot pass valve disease onto another person even if it has a rheumatic origin or if you had infective endocarditis.

Q: Is valve disease hereditary?

Valve disease is common so relatives might have valve disease by coincidence. However relatives of people with a bicuspid aortic valve have about a 10% chance of also having a bicuspid valve.

It may be worth considering an echocardiogram to check for this. Some types of mitral prolapse may run in families but this is unusual.

Q: Is valve disease dangerous?

Provided it is treated appropriately with surgery when this is indicated the results are excellent. Valve disease may be dangerous if unrecognised and symptoms are allowed to progress without treatment.

This means that if you have unexplained breathlessness or chest pain or dizziness on exertion you should see your usual doctor to see whether further tests may be necessary.

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Chapter Three What are the signs?

WHAT ARE THE SIGNS?

How do I know if I have heart valve disease?

Most valve disease causes no obvious signs or symptoms. Often, the first tell-tale sign is 'silent' to the patient, but not to a doctor with a stethoscope.

It's a murmur that might be detected during a routine health check at your GP surgery, or at a special clinic for an illness not related to the heart.

But sometimes heart valve disease does make itself known with symptoms. These are the most common.

Breathlessness

Breathlessness on a normal everyday exertion like walking, not playing sport or using a gym. Typically, this starts when walking uphill (say on the approach to a building) and then becomes progressively worse.

There are simple tests you can do at home or work that will help doctors (cardiologists) to diagnose heart valve disease, and treat it early.

- How many stairs can you climb before you have to stop to catch your breath?
- Do you get breathless only on steep hills or on mild hills - or even on the flat?
- Does it happen at normal walking pace - or only if you hurry or carry a bag?

Breathlessness at night

Breathless at night. If valve disease progresses, you may become breathless at night, because more blood returns to the heart when you're lying down.

Your cardiologist will want to know if you sleep on more pillows than usual, or even sleep propped up or in a chair. You may even wake at night with severe breathlessness after slipping off the pillows.



Ankle swelling

Ankle swelling, especially if associated with progressive breathlessness.

Otherwise common causes include varicose veins, being overweight and normal fluid retention.



Chest tightness, Dizziness and Fatigue

Chest tightness or heaviness

Dizziness, or fainting after walking strenuously

Fatigue during or after exercise. Some patients notice fatigue rather than breathlessness or chest pain

It's true that fatigue can also be caused by anxiety, poor sleep or anaemia, but progressive fatigue after exertion often points to an underlying cardiac problem.



Symptom spotting

Symptom-spotting is quite an art, both for patients and cardiologists

The worst thing people can do is put breathlessness down to 'getting older,' or some other handy excuse

It may be tempting to change the way you do your everyday activities to try to minimise the symptoms – **DON'T**

Now, more than ever, you must be honest with yourself

When you think you have a symptom, or a change in a symptom, tell your cardiologist at once. No detail is too small

He or she is already on his or her guard to spot the earliest symptom

You must work together to get the best outcome for YOU

When there are no obvious symptoms, it's down to **YOU** and your knowledge of your own body to own up to changes, not overlook them

Getting treatment

If your symptoms are obvious, you will probably be told that surgery is the only cure for your heart valve disease. It may be needed urgently, or in some weeks or months.

Try not to worry if this is the case. You can read the personal experiences of patients like you who have been told that surgery is the only treatment on *page xx*.

All surgery has some risks, especially if you have other medical conditions.

Your team of doctors will spend as much time as you need to discuss these risks. The first-hand accounts of other patients whose lives have returned to normal, or better than normal, after surgery should help to reassure you.

If you do not have symptoms, or your symptoms are slight, treatment is usually by observation. This is called **watchful waiting**.

"An important 'catch' for the unwary cardiologist is that people often slow down to avoid getting symptoms. With treadmill or bicycle testing there is no room for manoeuvre.

In people with severe aortic stenosis up to one half who believe they have no symptoms develop abnormal breathlessness or chest tightness on treadmill exercise.

This is why exercise testing is part of the routine assessment of any person who has severe valve disease but no apparent symptoms."

Professor Chambers

Key questions a cardiologist should ask

Cardiologists should always tease out a slowing down as an early symptom by questions like these:

- Is there anything you have had to give up?
- Can you walk as far and as fast as this time last year?

Often, a friend or partner is better able to notice – and report – a gradual slowing. People can be aware of a gradual slowing but assume that it is a normal effect of aging. Another person's first-hand opinion can be extremely useful in clinic, and should be encouraged.

Of course slowing down may be just that and has many causes other than heart disease. Painful or stiff joints, lack of fitness and weightgain could all be reasons for doing less. This raises the question of whether there should be any sort of 'screening' for heart valve disease.



It would actually be counter-productive, because it would find thickening or trivial regurgitation in about 44% of people aged over 65. The majority of these findings will never progress to more severe valve disease, and so the risk of causing unhealthy anxiety is not recommended.

What tests should you expect

Stethoscope

People aged over 65 should ask their GP to listen to their hearts with a stethoscope at well-person checks or when attending for flu-vaccination. This instrument that doctors usually wear around their necks can pick up the sounds of a valve or valves that are not working properly. This sound is called a murmur.

Echocardiogram

If there is a murmur, an echocardiogram is usually worth having. Echocardiograms should also be considered in people with lung disease who are more breathless than expected.

You should have an echocardiogram if you have

- atrial fibrillation (AF) because about 10% of people with AF also have valve disease
- Marfan syndrome because you are more likely than others to have mitral valve prolapse
- or if a relative has a bicuspid aortic valve, because you have a 10% chance of also having a bicuspid valve.