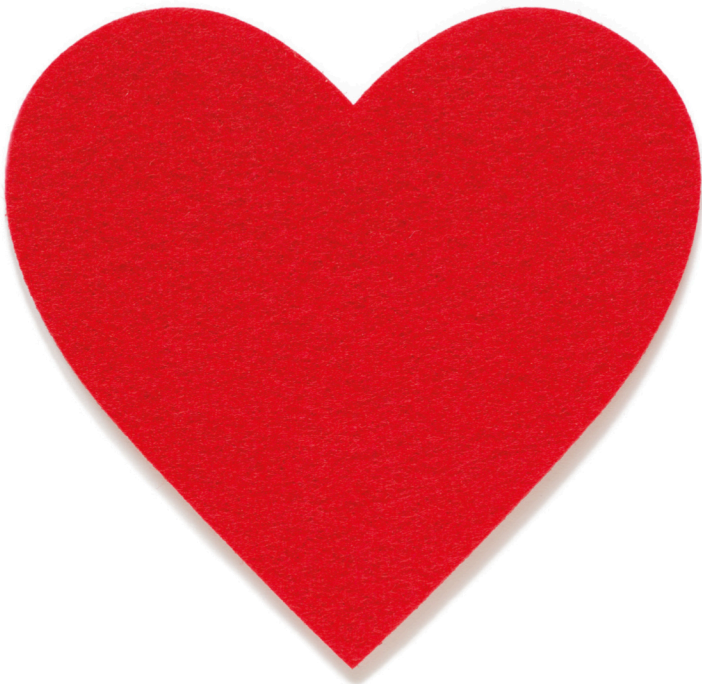


Marfan Syndrome: A Guide to Heart Surgery for Marfan Patients



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MARFAN SYNDROME

What is it?

Marfan syndrome (MFS) is an inherited genetic disorder of the body's connective tissue that affects both boys and girls of any race or ethnic group. It was first described by a French paediatrician, Dr Antoine Marfan, in 1896.

MFS can affect the cardiovascular system, eyes, lungs, gut, nervous system, and skeleton. Connective tissue, which helps provide structure to the body, binding skin to muscle, muscle to bone, is made of fine fibres and 'glue' called fibrillin. This tissue provides the stretchy strength of tendons and ligaments around joints and in blood vessel walls. It is also important in the eyes, lungs, and gut.

In MFS, fibrillin is deficient in connective tissue throughout the body, accounting for the unusual stretchiness and weakness of tissues.

Symptoms can differ widely from person to person with people experiencing mild to severe disability. Approximately 50% of people with MFS remain undiagnosed.

- In 75% of cases, it is an inherited condition due to a genetic change that is passed on from parent to child; 25% of cases are the result of a spontaneous genetic change (i.e. they are the first person in their family to have MFS).
- Any child of an affected parent has a 50% chance of inheriting MFS.
- It is caused by a change in the gene for fibrillin-1 on chromosome 15.
- MFS is rare, occurring in approximately 1 in 5,000 people worldwide. This would suggest there are around 15,000-20,000 people with MFS in the UK.
- Every year, on average in the UK there are over 200 new cases of Marfan Syndrome diagnosed.

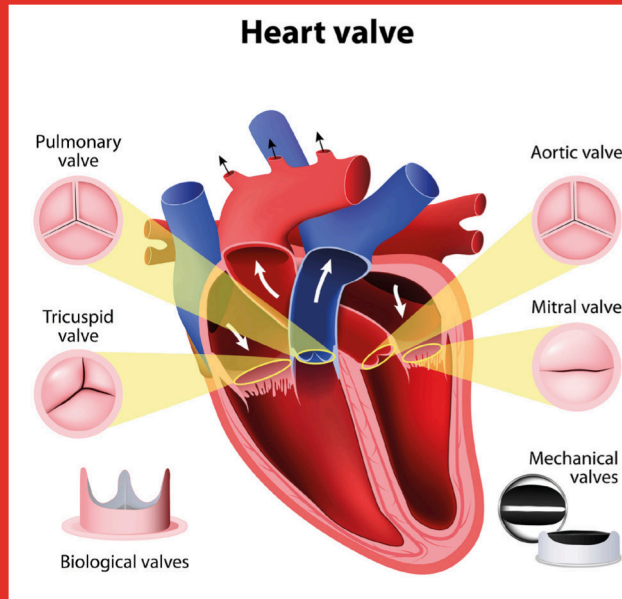
Introduction

If you have been diagnosed with Marfan syndrome, you are probably familiar with your Cardiologist who prescribes medication and invites you for regular scans and reviews to ensure that your heart and aorta remain healthy.

The main things a Cardiologist will be looking at are:

- To check the function of the heart valves to ensure these are not leaky (regurgitant); this can increase the workload of the heart as it has to deal with larger volumes of blood due to back flow from a leaking valve. This picture shows the position of the four heart valves. Individuals with Marfan syndrome can often have issues with a leaky mitral valve. If the aortic root is dilated, this can begin to affect the function of the aortic valve as it sits at the bottom of the aorta.

- To check the appearance of the aorta (the main blood vessel carrying blood from the heart to the rest of the body), particularly the aortic root and the ascending aorta to make sure that this is not enlarging and dilating to the point at which there is a significantly increased risk of aortic dissection (an emergency situation when there is a tear in the wall of the aorta): the picture of the aorta includes labels as there are certain regions of the aorta that your surgeon may discuss when planning an operation. You can use this picture in your appointment as a guide to help understand what is planned.



■ Regular follow-ups are important because any changes to your heart health can be picked up and managed appropriately. Although not always possible, a planned operation is safer. If your Cardiologist feels that you are reaching the point at which an operation may be necessary, they will refer you to a Cardiothoracic Surgeon for further investigations and discussion. When you meet your surgeon, he/she/they will be able to give you a detailed explanation about the best options for you and the possible risks and benefits of the surgery. Everyone is individual and therefore has a different risk for surgery. Your surgeon will look at all your information and be able to talk about this with you specifically.

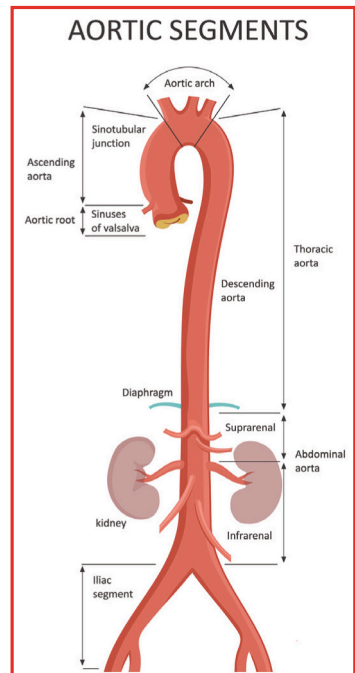
This booklet aims to provide you with some information about what to expect if you are referred for an operation. Your hospital will also provide you with resources to read. It can all feel overwhelming, but the Marfan Trust can answer general questions about surgery and most hospitals will have a dedicated person you can contact in the run-up to your operation.

This diagram of the aorta can help you clarify the location of any aortic dilatation or aneurysm you may have and visualise the area of the aorta that your surgeon is planning to treat.

There are many different types of heart surgery that you may undergo as an individual with Marfan syndrome, and it won't be possible to go into depth about all of these, but your surgeon will explain in detail the pros and cons of different types of surgery so you can make an informed decision with their guidance and support.

Some of the common operations you may be having are:

- Aortic root repair with aortic valve replacement
- Valve sparing aortic root replacement
- Ascending aortic aneurysm replacement
- Personalised External Aortic Root Support (PEARS)
- Mitral valve repair or replacement



Preparing for an operation

Once you know you are having a heart operation, it's important to try to remain as healthy as possible or make some positive changes to improve your general health:



■ **If you smoke** – stop as soon as you can. You can speak to your GP or pharmacist about local smoking cessation support. Smoking can increase the risk of complications after your operation, particularly with breathing.

■ **Try to maintain a healthy weight** – being overweight can increase the risk of complications following your operation so it will be beneficial if you can develop healthy eating habits prior to your surgery. You can request a dietician referral if you are struggling with this.



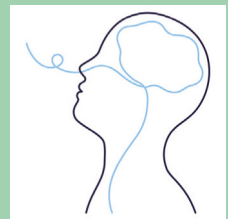
■ **Alcohol** – do not drink alcohol to excess. In the UK the recommendation is for both men and women not to drink more than 14 units of alcohol per week. Ideally, you should have at least a couple of alcohol-free days each week.

■ **Take regular exercise** – your ability to exercise will need to be guided by your medical team and they will explain what you are able to do. Keeping yourself as active as you are able will help with your recovery.



■ **Visit the dentist** – this is particularly important if you are having a valve operation. Infection within the teeth or gums can get into the bloodstream and cause an infection called 'endocarditis' which is more common after a valve has been replaced. It is also important to mention any crowns, dental implants or false teeth to your anaesthetist as they will need to be aware of these when inserting the breathing tube that will be in place for the operation.

■ **Deep breathing exercises** – it can be helpful to start practising taking deep breaths prior to your operation as this is very important once you have had surgery. Take deep breaths whilst sitting upright, hold the breath for a count of three before breathing out, and repeat three or four times during the day. It's important to expand your lungs as much as possible during these breathing exercises.



Patient Perspective

Frances suffered from severe needle phobia that delayed her diagnosis of Marfan syndrome due to her struggle with having a blood test for genetic screening. She was very anxious about the prospect of heart surgery.

Frances discussed these issues with her GP and her surgical team, and they were able to come up with ways to help. In her case, she was introduced to a numbing cream that can be applied prior to the insertion of any needles/ cannulae and she was prescribed medication to help with her anxiety in the build-up to the operation.

She would encourage people to talk to their clinicians about their fears and worries as they will be able to help and offer possible solutions.

Pre-assessment visit

Once you are on the waiting list for an operation, you will be invited to the hospital for a 'pre-assessment visit'. This will usually be with a nurse specialist who will arrange various tests for you and have a conversation about what to expect.

The appointment will often take several hours including all the tests, so be prepared to be at the hospital for some time.

The tests you have could include:

- Chest x-ray
- Blood tests
- Electrocardiogram (ECG)
- Echocardiogram (Echo)
- Lung function tests
- Doppler scans of the neck vessels
- Swabs of your nose/throat/groin

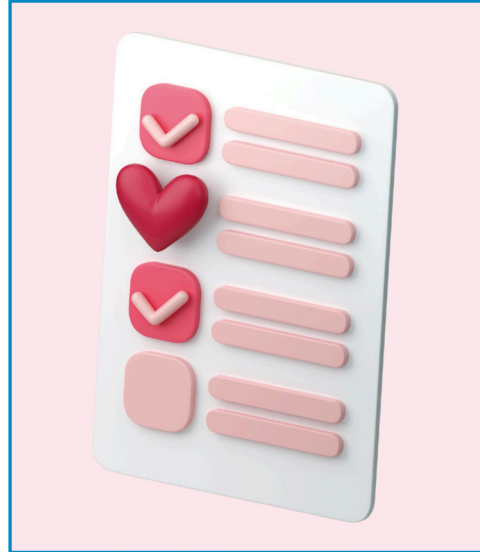
You may also require an extra CT or MRI scan to give detailed imaging for the surgeons to plan your operation. For the PEARS procedure a CT scan needs to be done for your personalised aortic root support to be created. These scans probably won't be done on the same day as your pre-assessment visit but in the weeks prior.

All these tests give the team important information about your general health and make sure you are prepared for the operation. You may also see an anaesthetist at this point so they can make sure your anaesthetic during the operation can be given safely.

How to prepare for your pre-assessment visit

It is helpful if you can bring someone along to your appointment. Lots of information will be shared with you and it can be useful to have someone else listening too. Feel free to take notes.

- ✔ **Write a list of questions before you go to the hospital.**
- ✔ **Bring a list of all your medications and doses.**
- ✔ **Let the team know about any allergies you have.**
- ✔ **Think about any special adaptations you might need. For example, if you are very tall they may need to arrange a special hospital bed for you prior to admission.**
- ✔ **Have a think about your plans when you go home – ideally you need someone to stay with you for at least a week after you are discharged from hospital, and you will need help with shopping/cooking, etc.**



In particular, ask yourself these questions:

Do you have someone who can collect you from hospital?

Do you have someone living with you?

Can someone stay with you after your operation?

Can you stay with a friend/relative?

Patient Perspectives

■ **Francesca** is currently under review by her cardiac team and her surgery is being planned. Francesca is a young woman with MFS; she is the only one in her family with MFS. She feels it is important to bring along a trusted friend, relative or partner to appointments. This person may often ask different questions, perhaps things that you hadn't thought of or might find difficult to bring up.

Planning for surgery is anxiety-provoking and it's important to be physically and mentally prepared for any operation. Francesca was finding balancing work difficult and decided to talk to a trusted colleague about her concerns, which she found very beneficial. She has also reached out to other people with MFS who have been through surgery. As the only person in her family with MFS, she connected with a friend who was able to give her lots of useful advice and reassurance.



■ **Rebekah** had mitral valve surgery when she was 21 years old. In preparation for her surgery, she had several appointments with her surgeon and cardiac team. She was given lots of opportunity to discuss all aspects of her surgery including important considerations like possible future pregnancies. Rebekah always wrote down questions she wanted to ask at the appointment and would go along prepared.

Some people can feel slightly intimidated by the 'experts'; however, Rebekah points out that as an individual with MFS she has had regular interactions with medical professionals throughout her life due to multiple surgeries (particularly to her eyes) so feels that it is quite normal to sit down with the medical team, her parents and her partner to ask questions.

Admission to hospital

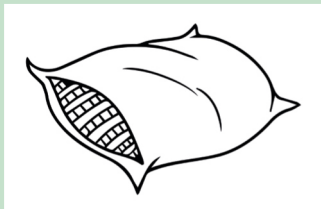
The hospital will let you know when your operation is scheduled and explain the admission process to you. Some hospitals admit patients the evening before their operation and some on the morning of their surgery. Unfortunately, it is always possible that **your operation may be postponed** due to emergencies.

What to bring

- All your medications
- Loose-fitting, comfortable clothes. Front-fastening tops are recommended as these are easier to get on and off with your surgical wound
- Women should bring well-fitting bras (without underwiring); ideally these should be front-fastening for ease or bring tops with secret support panels
- Slippers/comfortable house shoes that cover the whole foot
- Toiletries
- Hearing aids/glasses/dentures

Patient Perspective

Rebekah recommends button-up pyjamas and tops for the duration of the hospital stay; with a chest wound it is much easier to get dressed and undressed. Bring a large, comfortable pillow that can help with getting into a comfortable position following surgery. (You may need a relative to bring this in for you once you are back on the ward as there will be limited space for storage and you won't be able to move around in bed that much immediately post-operatively.)



You may be asked to have a shower with a special soap the evening before and on the morning of your operation – the nursing staff or pre-assessment team will explain this to you.

You will not be able to eat or drink for at least six hours prior to your operation. You may be able to have clear fluids up to two hours before, but the hospital will let you know and ensure you understand what you can and cannot do.

Before your operation, the medical team will ensure you have signed your consent form. You may have done this during your pre-assessment visit if you saw your surgeon then. The doctors will check through this with you again and if you have any additional questions, do ask.

Anaesthetic

Once it is time for your operation, you will be taken to the operating theatres and brought to the anaesthetic room. You may walk down or arrive on a bed.

The anaesthetist is the doctor responsible for your breathing, blood pressure, heart rate, kidney function, temperature and blood volume throughout the operation and in intensive care. They therefore need several monitors to enable them to do this. You will be attached to a heart monitor and have a probe on your finger to monitor your oxygen levels. A small cannula (plastic tube) will be inserted in an artery in your wrist and in a vein. Once these are in place, you can be given anaesthetic medication at which point you will go to sleep.

There are lots of different types of heart surgery and this booklet doesn't go into detail about all of these. Your hospital and doctor will provide specific information about the type of operation you will be having.

Immediately post-operatively

When you first wake up following your operation, you will be in a specialist recovery area which is like an intensive treatment unit (ITU). You will still be very sleepy and attached to various machines and monitors:

■ A breathing tube in your windpipe will be attached to a ventilator to help you breathe while you are asleep. Once the medical team is happy that your condition is stable, and you are awake enough to breathe for yourself, this tube will be removed and you will be encouraged to take some deep breaths and start to cough if you need to. You will have an oxygen mask over your mouth and nose.

■ Drips in your neck, and in your arms: some of these are to monitor your blood pressure and some are to give you medication and fluid. These will usually be removed over the first 24-48 hours once you are eating and drinking for yourself and can take tablets again.



- A catheter in your bladder collecting your urine. This will also usually be removed in the first 24-48 hours.
- Chest drains are tubes placed around the heart and lungs during the operation to drain any blood or fluid from the area after the operation. These will be removed once there is no further drainage.
- Pacing wires are placed around the heart and can be used if there is any disturbance to the heart rhythm after the operation. These will also be removed a few days after the operation.
- A heart monitor will be attached with stickers on your chest.

Patient Perspective

Many of the patients who helped with this leaflet mentioned that this first period of their recovery was much more difficult for their family than for them. The person having the operation may have very little memory of their stay in ITU while their loved ones and family remember this clearly and can find it difficult to see their loved one vulnerable and attached to so many monitors and machines.

It's important that your family understand what to expect. Usually there will be a limit on the number of people who can visit recovery and the length of time they can stay, so it's helpful to nominate a family member to disseminate information to other family and friends.

Most people wake up a few hours after their operation but there may be reasons that the medical team decide to keep you asleep for longer and this is normal.

Most patients who have had heart surgery will have a **sternotomy scar** down the centre of the chest; the breastbone is split to allow access to the heart. Wires and stitches are used to close the bone and tissues to allow them to heal and it is important that during your early recovery you **do not try to do heavy pushing/pulling or lifting** to give your breastbone time to heal.

Some patients who have aortic surgery will instead have a **thoracotomy scar** from the middle of the shoulder blades at the back, down and across the ribs on the left-hand side. The physiotherapists and nurses will show you how to support your chest wound when you cough and do deep breathing exercises and you will be given a pad to support your wound when you do this (a rolled-up towel or cushion is often sufficient for this). They will also show you **how to get in and out of bed comfortably and safely**.

Pain relief

It is natural to be concerned about pain following an operation. Some discomfort is inevitable, but **it is important that you are not in lots of pain after your surgery**. At first, you will be given strong pain relief through the drips. Once you can take tablets again, the nursing staff will give you regular medication. If you are in pain, you will be less likely to take deep breaths, cough and get up and about. All these things are very important for your recovery, so do **let your medical team know if you are uncomfortable** as they can adjust your medications. Taking regular pain medication helps to keep you comfortable. **Don't wait for the pain – take your medications regularly to stay on top of it.**

Constipation

A side effect of some pain medications, along with possible dehydration and your reduced mobility straight after the operation, mean that you can become constipated. In the first days after your operation, the medical team can prescribe medication to help you go to the toilet and they will ask you about whether you are struggling with this. Do let them know if it's an issue as you can become uncomfortable and need to **avoid straining excessively after surgery**. Once you are more mobile, eating and drinking a normal diet and taking less strong painkillers, this should resolve.

Patient Perspective



Gareth underwent two surgeries in fairly quick succession in 2016/7. One was an emergency operation due to a Type B aortic dissection and the second followed 12 months later due to further dilatation of the aorta at a different point. "My top tips are to take recovery one day at a time, stay on top of the pain and look back to how far you have come since surgery, rather than comparing yourself to how you were before. It's like becoming a toddler again and having to slowly build up the stamina, strength and energy of an adult. It takes time and can't be achieved overnight.

"I wasn't offered cardiac rehab, but I think I would have benefited from it. I started to beat myself up for not making faster progress, until my cardiologist said to me, 'Gareth, it takes people 18 months to properly recover from what you've had.' She lifted a weight from my shoulders that day.

"In consultation with my GP, who is extremely supportive, it became clear that I would not be returning to work. That was a big adjustment, but I realise now, seven years on, that it was good advice and the right decision. I have accepted and adjusted to my 'new normal'. Every patient is different, but I hope that sharing my experience will help others on their journey."

Back on the ward

Getting up and about as quickly as possible after your operation is important. This helps to prevent complications such as chest infections, constipation and pressure sores.

- At first you will just move from your bed to your chair with some guidance from the nurses who will show you **how to move and change positions safely**.
- You will be seen by the physiotherapists who will give you advice about **coughing and deep breathing to aid your recovery**.
- After a day or two, you will be able to **walk for short distances** and can aim to increase this a little bit each day.
- Before you go home, the physiotherapist will make sure you can go **up and down a flight of stairs safely**.
- It's important to **keep up this regular walking and exercise once you get home**.

Patient Perspective

Frances underwent a PEARS procedure in 2020 (Personalised External Aortic Root Support). She already had a diagnosis of Marfan syndrome and had unfortunately lost her sister to aortic dissection, so she thoroughly researched her options for surgery and asked her GP to refer her to one of the aortic surgeons in the UK performing the PEARS procedure. Remember, patients can investigate their options and should feel able to ask for second opinions if they feel this is necessary.

Frances had some complications and some comorbidities that meant her recovery took slightly longer and she was not discharged until day nine following surgery, but she accepted that everyone recovers differently, and the hospital kept her in until it was safe for her to go home.

She took her recovery slowly and listened to her body while gradually trying to increase her activity levels. She was supplied with an app to carry out her Cardiac Rehab due to the COVID pandemic but found this quite useful as she was able to follow it at her own pace when she felt well enough to do the recommended exercises.



Washing and dressing

Once you are back on the ward, you will be able to put on your own clothes again. You will be encouraged to get back to your usual routine for personal hygiene so if you usually have a shower, you will be able to do this. **The nurses will advise you on caring for your surgical wounds** which may differ depending on the type of operation you have.

Eating and drinking

Straight after your operation, you will be getting fluid through a drip. Once this is removed, **it is important that you drink lots of fluids to remain hydrated.**

It is normal for your appetite to be reduced after an operation and your sense of taste or smell might be affected for a while. However, it is important to start eating again as your body needs nutrients to heal following surgery. You will be given regular meals and snacks in hospital and **need to keep up good nutritional intake once you get home** too. Prior to your admission to hospital, it can be helpful to batch-cook and freeze some nutritional meals that you can eat when you get home with minimal effort.

Some people can struggle with nausea or constipation following surgery. The nurses will talk to you about this to make sure it's not a problem for you. Medication can be given to help with this if necessary.

Patient Perspectives

Darren underwent aortic root replacement and aortic valve repair in 1985 and then in 2021 had redo aortic root replacement as well as aortic and mitral valve repair. He offers a list of helpful tips developed from his experience of surgery and recovery:

- **You need to be mentally as well as physically strong to recover:**
- Don't suffer with things that are worrying you – speak to the nurses or doctors if anything is bothering you and they will be able to help.



- **Take some time to think** about how you felt on the previous day and how you have improved since then, even in little ways.
- **Take time to speak to the nurses and other patients** – it will lift your mood and help pass the time.
- **Spend some time with loved ones**, both while in hospital and once you are back at home. They will help improve your mood and motivate you to get better.
- **Try to be positive** – you will have bad days, but try to put negatives to the back of your mind until you're well enough to deal with them.
- **Listen to the doctors** – they're the experts and want the best for you.
- Physiotherapists – they seem like a pain(!) but are so **very important in getting you moving again**.
- **Keep moving while in bed**, whether it's just lifting your legs and raising your arms – this helps prevent possible complications and helps the body recover.
- **Keep your energy up** and don't forget to snack between meals, whether it's a banana, slice of cake or a few biscuits. The body needs this to recover.
- Pain – you will get pain, but keep on top of it and you will be able to do more and feel better both mentally and physically. **Taking painkillers is not a sign of weakness.**

Patient Perspective

Martyn has had numerous surgeries, including cardiac, and he also has some useful insights into recovering after such a big operation:

- **Don't be afraid to ask for help.** Speak to your friends and family – they will most likely be keen to help you after your operation, so reach out to them and let them know what you need.
- **Speak to your GP before you go into hospital** if you think you will need a care package after you go home (for example, if you live alone). This may be something you are worrying about, and your doctor will be able to help plan how to go about organising this.
- **Bring several pairs of pyjamas and your own toiletries** to hospital. You feel much better if you have your own things.
- **Have a tablet device or laptop to help pass the time** following your surgery (you may need to ask the ward about when it would be best to bring this in as they may not be able to look after it while you are in surgery and ITU).
- Martyn suffered from **vivid dreams and some hallucinations** straight after his operation which he found frightening; this can be quite common after heart surgery. His advice is to talk this through with the doctors and nurses so they are aware and can help reduce the anxiety surrounding this.
- Having loved ones to visit is wonderful but tiring! Again, **don't be afraid to tell your visitors that you are tired and need to rest.** Perhaps limit to just one visitor a day for the first few days after your operation.

Going home

Most people are ready to go home around five to seven days after their operation, but everyone is different, and **the hospital won't send you home until they feel you are fit and well.**

Before going home, you will see a nurse or physiotherapist from the Cardiac Rehabilitation team who will be able to give you advice about increasing your activity levels when you go home and will enrol you in the cardiac rehabilitation classes that usually start around six weeks after your operation.

These classes usually happen twice a week and include exercise and advice about **healthy lifestyle, protecting your heart health in the future and managing stress.** They also give you the opportunity to meet other people in your area who have recently had heart surgery.

Patient Perspective

Many of the patients who contributed to this leaflet felt that the cardiac rehabilitation classes were often targeted more at individuals who have had coronary artery bypass surgery, so were not always aimed at their specific needs. However, they all found the classes helpful in restoring their confidence to exercise and understanding how to safely reintroduce exercise to their lives. If they did have specific limitations due to their Marfan syndrome, the rehab team were able to make adaptations or modify exercises. Please discuss these with the nurses or physiotherapists.

The nurses on the ward will make sure you have all the medications you need to take home and will explain how to take them. They will also provide a list of these for your GP as well as information about the surgery you have had in your Discharge Summary.

It's important to make an appointment to see your GP in the first couple of weeks after your operation as your medicines may have changed and they will need to update your prescription and be aware of the operation you have had.

You may have one or two remaining stitches (for example at the site of your chest drains or pacing wires) and the nurses will explain when these need to be removed; the practice nurse at your GP surgery will be able to do this for you. You will also be given a follow-up appointment with your surgeon; this usually happens six to eight weeks after your operation. At this appointment they can check that

all your surgical wounds have healed well, that you are taking all the correct medications and that you are not experiencing any troubling symptoms.

It is important to remember that many patients with Marfan syndrome will have had surgery to the aorta and this needs long-term follow-up. Lots of hospitals that do aortic surgery will have specialist clinics to monitor patients, so ask your doctor how this will be done and how often you will need to have scans and appointments. Make a note of when your follow-ups are due so that if you are not offered an appointment you can get in touch with the medical team.

Moods, emotions, rest and relaxation



After a big operation **it is normal for your emotions to be up and down for a while.** You may have days when you feel elated and relieved that the operation is over and you may have other days when you feel overwhelmed, irritable or tearful. This is normal and should settle down within the first couple of months after your operation.

Make sure that your family and loved ones understand that this is a normal part of your recovery. If you continue to feel like this long-term or begin to feel depressed, then do get in touch with your GP.

Your sleep patterns may be disturbed after your surgery and you may have vivid dreams. Again, this should settle down within the first couple of months. You will be more tired after the operation and you need to take extra rest while remembering to stay active and commit to gradually increasing your daily activity.

You will need to sleep on your back for the first few weeks following surgery to allow the sternum to heal. It can be more comfortable to sleep propped up with some pillows, so do experiment and find a position that is comfortable for you to get quality rest and sleep.

Pain

You will be sent home with some pain relief, and it is important to keep taking this regularly when you first leave hospital. If you are in pain, you will be less likely to get up and move around, which is important. Once you start to feel more comfortable you can drop a dose of the painkiller, perhaps in the middle of the day to see how you feel.

Patient Perspective

Karen underwent urgent surgery in 2021 during the COVID pandemic which meant her prolonged hospital stay was made more difficult due to no visits from her family.

She had a mechanical aortic valve and replacement of the aortic root and ascending aorta. Her Marfan syndrome wasn't diagnosed until after her surgery. Karen shares many things she found helpful during her prolonged hospital stay:

- When you have appointments with your medical team or surgeon, write down your questions and their answers. You may not take in lots of the information at the time, so it's useful to have this noted down.
- Make sure you have things with you in hospital to keep you occupied: books, notebook and pen, phone, tablet device with headphones.
- Work with the physio and follow their advice regards exercise: marching on the spot, moving around the ward and breathing exercises all help with your recovery.
- Bring front-fastening tops or large, stretchy sports tops that are easy to get on and off. Sports bras are a good option post-op.
- Take rest when you feel tired. At first, just sitting out of bed for a few hours can feel exhausting and you shouldn't feel bad about having a sleep. Once you get home, you will still get tired and sometimes that fatigue can feel overwhelming. Listen to your body and take rest when you need it.
- Karen found it much more comfortable to sleep in an upright position in the early days following her operation, so she invested in some comfortable pillows, including a triangular pillow to help her get the quality sleep she needed to recover.
- The number of medications you come home with can feel overwhelming. The number of tablets will probably reduce over time, but it's important to take time to get these organised, so you are taking the right tablets at the right time.
- There are good days and bad days after surgery and this is normal. You can feel tearful and low. It can be helpful to keep a note of when you have a 'bad' day so you will hopefully see these reducing over time.
- Cardiac rehab was via phone and video due to COVID. However, once the restrictions were eased, Karen asked to be re-referred and joined an in-person class which she found very useful especially for guidance about how much you can push yourself.
- Going back to work can be incredibly tiring, so make sure you have spoken to your employer and arranged a phased return which can be reviewed on a regular basis.

Activity

The aim is for you to be feeling back to your baseline (how you felt before your operation) or better by about 12 weeks after your surgery. You should hopefully be getting to the point at which you can return to 'normal' activities and work. However, everyone is different and everyone's job is different. For example, a builder will not be able to go back to work lifting and carrying 12 weeks after surgery while an office worker may be able to begin a phased return at that point. It is important to have a conversation with your employer or your school/university/college, so they have plans in place to support your recovery.

Timelines are not always helpful and depending on the type of surgery, other illnesses or effects of Marfan syndrome, many patients may need much longer to recover and return to 'normal life'. In some instances, people decide to adapt their work life or career choices permanently following surgery.

Individuals with Marfan syndrome have a range of different affects and symptoms and will therefore need to tailor their recovery to their specific needs. The Cardiac Rehabilitation team will also be able to help with this.

When you have had heart surgery, it is important to maintain a healthy lifestyle to try to keep up any positive changes you have made:

- No smoking
- Maintaining a healthy weight
- Taking regular exercise
- Paying attention to your mental health and wellbeing
- Keep going to the dentist regularly (at least every six months); if you need antibiotic cover for dental treatment, your surgical team should have told you about this and will have given you a card to show your dentist.

Sexual relations

You can **resume sex once you feel that you have recovered from your surgery**. You can treat this like other forms of exercise and physical activity that you gradually build up over time. Do not choose a position that would put extra weight or pressure on your chest wound. **Talk to your partner as they may feel anxious as well**. You may not feel physically or mentally ready for a while and this is OK.

Some of the medications that are usually prescribed after heart surgery can interfere with the ability to achieve and maintain an erection. If this is an issue, talk to your GP. You also need to **consult your GP before taking any medication like Viagra®** as this may also interfere with some of your medications and not be suitable after your surgery.

Warfarin

Following some types of cardiac surgery, particularly replacement of a heart valve with a mechanical valve, **patients will need to take lifelong warfarin**. Warfarin is a blood thinner and its function in this situation is to prevent blood clots forming on the surface of the replacement valve which can detach and enter the blood stream, potentially causing a stroke.

You will be referred to an 'anticoagulation clinic' before you go home, and they will be responsible for performing **regular blood tests to ensure your blood 'thickness' remains within the target range** set by your surgeon.

Blood thickness is measured using **International Normalised Ratio (INR): normal blood has an INR around 1.0**. It is important that you know your target INR range. It will be documented in your medical records and in your yellow anticoagulation record book which will be provided to you before you go home.

Sometimes it can take a while for your INR to settle down and you may need quite regular blood tests, but over time this should become more predictable. **The clinic will recommend how regularly you need to have the blood tests.**

There is now the possibility of home testing kits for checking INR. At present, the machines need to be purchased privately by patients and the disposable parts of the test kits need to be supplied by the GP. This needs to be discussed with the anticoagulation clinic or GP practice to see if they think this would be suitable or appropriate for you. This is always done in partnership with your anticoagulation service but can make life easier if you are busy and find it difficult to get to frequent appointments for blood tests.

Some food, drinks and medications can increase or decrease the effect of warfarin so before you start any new medication, even over-the-counter remedies, do mention to your pharmacist that you are taking warfarin. Some people choose to wear a medical alert bracelet.

It is also important to let other doctors or medical professionals (including dentists) know that you are taking warfarin prior to any other procedures as adjustments may need to be made to reduce the risk of bleeding.

Dental work

If your aortic valve has been replaced, it is important to protect this from potential infection – dental hygiene is vital. **Clean your teeth twice a day and visit the dentist every six months**, including visits to the hygienist. Ensure that your dentist is aware that you have a heart valve replacement and if you take warfarin or an alternative blood-thinning medication.

Patients who have had a valve replacement will need **a single antibiotic dose an hour before any dental treatment** (not for check-ups). Your surgeon will let you know if this applies to you; it is important to obtain this information before you go home. The dentist can prescribe the required antibiotic for you.

Another route that infection can enter the bloodstream is via tattoos and piercings, so apart from basic ear-piercing the recommendation is to **avoid further tattoos and piercings if you have had a valve replaced**.

Endoscopy

It is also important to protect your aortic valve from infection when anticipating an endoscopy of either the colon or the urinary or genital tract. Colonoscopy has become quite a frequent investigation and a screen for those with a family history of polyps in the colon. Prophylactic antibiotics are essential for those with a replaced or a repaired valve.

Patient Perspective

Steven underwent aortic root replacement and tissue aortic valve replacement in 2004 when he was 26 years old and then unfortunately needed repeat surgery in 2006. He has been fit and healthy since then.

When Steven came to the point of requiring surgery, he had recently been referred to a local hospital and the team looking after him were new. He found this challenging as his previous team had known him well and were able to give him advice tailored to his Marfan syndrome and his personal situation. Sometimes, a lot of the advice about cardiac surgery was very specific to patients undergoing coronary artery bypass surgery, which is a different operation carried out for different reasons, and he found this frustrating.

Steven did find the cardiac rehab classes useful, but he was a very young man in a group of mostly elderly patients who were starting from a much lower level of fitness and had different needs to him. However, the classes gave him the confidence to start exercising in a safe environment again and provided guidelines for increasing this and getting back to his previous level of fitness, so

he would recommend them.

It was lonely in hospital. Steven did have long stays due to complications but again, due to his young age he was often the only young person on a ward of quite elderly patients. He advises having things to keep you amused and to pass the time. One thing he did was speak to medical and nursing students about Marfan syndrome, a great opportunity for them to learn about the condition by seeing it first-hand and the serious implications it can have for those with the condition.

Driving

While you do not need to inform the DVLA after heart surgery, **you shouldn't drive for at least one month**. You may need to check with your insurer as they could have different rules. **Other rules apply if you hold an HGV license**, so you need to check this on the DVLA website. **Make sure you don't drive until YOU feel ready**. Remember: everyone recovers differently.

Prior to surgery, you must tell the DVLA if you have Marfan syndrome. You may be prosecuted if you are involved in an accident as a result. Different forms are available for car or motorcycle licence, and bus, coach, or lorry licence. In addition, you must tell the DVLA if you have an aortic aneurysm. Ask your doctor or consultant if you are not sure.

Symptoms to be aware of

Symptoms to be aware of in the weeks following your surgery:

- Redness, increased pain, heat, swelling or pus around your wound
- Fevers or night sweats
- New or severe palpitations
- Repeated dizzy spells or blackout
- Chest pain
- Becoming very short of breath



These symptoms need further investigation, and you should alert your GP or the hospital's advice line (if they gave you contact details). If you become very unwell or these symptoms are severe, you should seek urgent medical attention by attending A&E or calling 999.

Follow-up and emergencies

Many individuals with Marfan syndrome will be having specialist surgery that requires referral to a specialist centre a significant distance from home. You will have a follow-up appointment with your surgical team, but it is possible that you will then be referred to your local team to continue your ongoing follow-up and surveillance. If you become acutely unwell and need an ambulance, you will be taken to your local A&E department. It is important that you have some information about your operation and medical history available.

Some patients choose to upload a photo of their latest clinic letter or discharge summary to the Medical ID page on their mobile phone; alternatively keep a paper copy in your wallet or bag. It is helpful if you can provide some information to the medical team:

- What operation you had
- When you had your operation
- Contact details for your specialist team
- List of your current medications

If you are unwell, you may struggle to recall all this information, so having it stored somewhere on your phone makes it much easier for you and the emergency team treating you. If you have had recent surgery, they will probably be keen to get in touch with your surgical team and this information can expedite the process.

Useful links

■ **British Heart Foundation – Heart Surgery Leaflet**

<https://www.bhf.org.uk/information-support/publications/heart-conditions/having-heart-surgery>

■ **British Heart Foundation – Cardiac Rehabilitation Information**

<https://www.bhf.org.uk/information-support/support/practical-support/cardiac-rehabilitation>

■ **Aortic Dissection: The Patient Guide**

<https://tinyurl.com/ADPatientGuide>

■ **NHS Smoking Cessation Services**

<https://www.nhs.uk/live-well/quit-smoking/nhs-stop-smoking-services-help-you-quit/>

■ **Drinkaware – alcohol consumption**

<https://www.drinkaware.co.uk/facts/alcoholic-drinks-and-units/how-much-alcohol-is-too-much>

■ NHS Healthy Eating Guidelines

<https://www.nhs.uk/live-well/eat-well/food-guidelines-and-food-labels/>

■ DVLA Health Conditions Checker

<https://www.gov.uk/health-conditions-and-driving>

<https://www.gov.uk/guidance/cardiovascular-disorders-assessing-fitness-to-drive#marfan-syndrome-and-other-inherited-aortopathies>

About the Marfan Trust

Co-founded in 1988 to fund research into the cause and treatment of Marfan syndrome, the Marfan Trust is the sole charity in the United Kingdom dedicated to improving and saving the lives of people with the condition. It funds its own laboratory, named after a young doctor, Sonalee Wijetunge, who died during her ward round of complications from Marfan syndrome. The results of the laboratory's internationally recognized research enable doctors and surgeons to provide better treatment for patients in the short and long term.

The Trust is committed to its three main objectives:

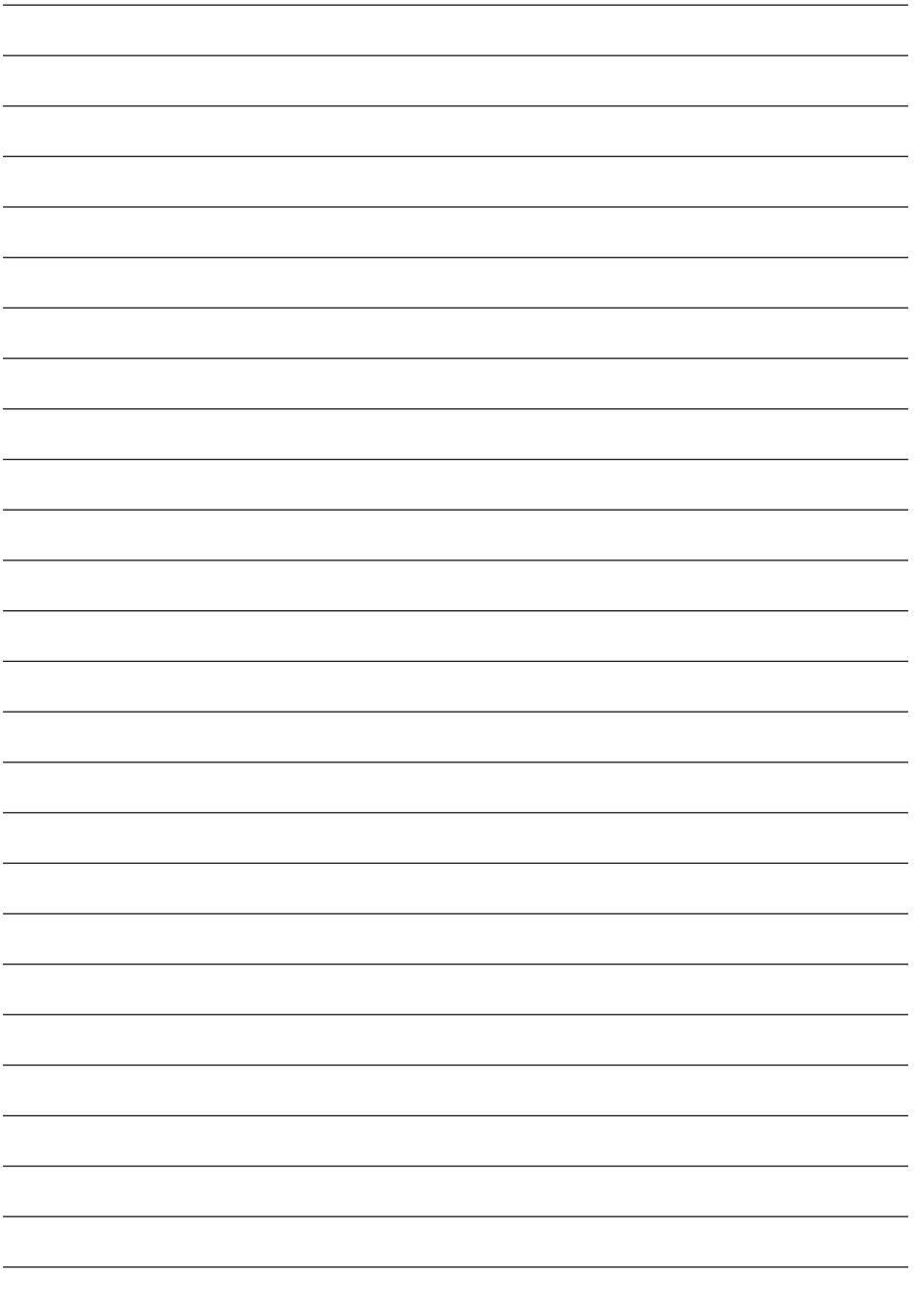
■ **Support:** the Trust provides personalized support and guidance to those with Marfan syndrome and their families through its bespoke medical helpline.

■ **Education:** the Trust is tireless in raising awareness of Marfan syndrome and all its aspects through educational literature, a strong social media presence and an information-filled website. It is estimated that approximately 18,000 people are living with Marfan syndrome in the UK, with many of these anticipated to be dangerously, and possibly life-threateningly, undiagnosed. By educating the medical profession and general public through pamphlets, publications, updates on cutting-edge research projects, and advice and guidance on living with Marfan syndrome, we hope to reach the undiagnosed while helping the diagnosed.

■ **Research:** the Trust undertakes cutting-edge research in its self-funded laboratory, easing the diagnosis and improving the treatment for people with the syndrome.



The Charity's cofounder, Dr Anne Child, MD FRCP, pioneered the international consortium that discovered the causative gene for Marfan syndrome and has since dedicated her career to securing a healthier, happier future for those with the condition. From setting up 22 national Marfan genetic clinics to discovering new genes behind lens dislocation and aortic aneurysms, she has made an indelible difference.



You can help to secure the Marfan Trust's future by becoming a member today for just £3 per month. Membership unlocks premium content published by the Trust and allows you to keep up with all things Marfan-related and be part of a supportive community.



You can also donate to our charity via bank transfer for any amount:

BANK: Charities Aid Foundation (CAF)

ACCOUNT NAME: The Marfan Trust

SORT CODE: 40-52-40

ACCOUNT NUMBER: 00017677

REFERENCE: Your Name Here (plus campaign name if relevant)

Alternatively, you can contribute via:

Just Giving: <https://www.justgiving.com/marfantrust>

PayPal Giving: <https://www.paypal.com/gb/fundraiser/charity/23440>

■ By donating to the Marfan Trust you are contributing to an ever-growing body of knowledge on the condition, allowing more doctors and medical specialists to deliver the best possible treatment to patients affected by Marfan syndrome.

There are many other ways and means of supporting our charity and getting involved. Why not visit our shop, a trove of lovely Marfan-related things including Dr Child's book for children, "Jeremy Wins". Or help to fund a piece of equipment in Dr José Aragon-Martin's Sonalee Laboratory and have your name etched for posterity.

Thank you for supporting us and the work we do:
Marfan Trust Hotline
0333 011 5256



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