Marfan Trust
Supporting Research into Inherited Aortic and Related Disorders
Loeys Dietz
CONNECTING THE UNCONNECTED AND STRENGTHENING LIVES

Marfan Trust Guide to Heart Surgery

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What Are Marfan & Loeys-Dietz Syndromes?

Marfan and Loeys-Dietz syndromes are inherited genetic disorders of the body's connective tissue, affecting any gender, race or ethnic group. Connective tissue helps provide structure to the body, binding skin to muscle and muscle to bone. It also supports the internal organs.

Connective tissue is made of fine fibres and 'glue'. One fibre is called fibrillin. In Marfan syndrome (MFS), a change in the fibrillin-producing gene, fibrillin-1, means that this protein is deficient in connective tissue throughout the body, creating an unusual stretchiness and weakness of tissues. This has far-reaching implications and can affect the eyes, lungs, gut, nervous system, skeleton and, most dangerously, the cardiovascular system. Symptoms can vary widely from person to person with people experiencing mild to severe manifestations. MFS affects roughly 1 in 3,000 people which means that approximately 18,000 people in the UK have MFS. We estimate that half remain undiagnosed.

75% of patients inherit the condition whilst 25% develop it as a result of a spontaneous (new) gene change. Each child of an affected parent has a 50% chance of inheriting Marfan syndrome.

Every year, on average in the UK there are over 200 new cases of Marfan Syndrome diagnosed.

Marfan syndrome was first described by a French paediatrician, Dr Antoine Marfan, in 1896. It is caused by a change in the gene for fibrillin-1 on Chromosome 15.

Patients with Loeys-Dietz syndrome (LDS), were once thought to have Marfan syndrome, such are the similarities between the two conditions.

LDS was first described in 2005 by Drs Bart Loeys and Harry Dietz. It is caused by a variant in one of the genes in the transforming growth factor-beta signalling pathway, TGF-β. The genes that cause LDS are: TGFβR1, TGFβR2, SMAD3, TGFβ3 and TGFβ2. TGF-β is a crucial signalling pathway involved in various cellular processes, including cell growth and immune responses. It is a key player in development and tissue repair. When a gene change occurs, it has implications for many systems of the body. Whilst symptoms vary from patient to patient, with some being similar to those of Marfan syndrome, the most characteristic are: arterial tortuosity, aortic enlargement, bifid uvula, curvature of the spine, high-arched palate and over-crowded teeth.

This pamphlet is intended for patients with both Marfan and Loeys-Dietz syndromes.

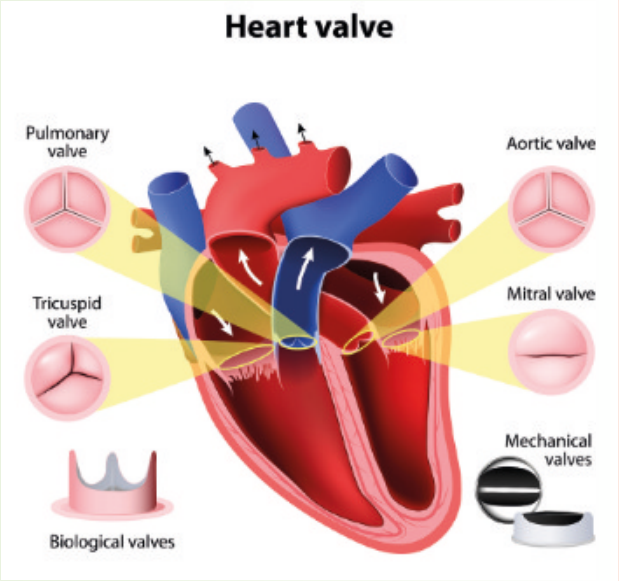
Loeys-Dietz syndrome is much rarer than MFS and the incidence is currently gauged to be 1 in 100,000 although this is probably an underestimation.

Introduction

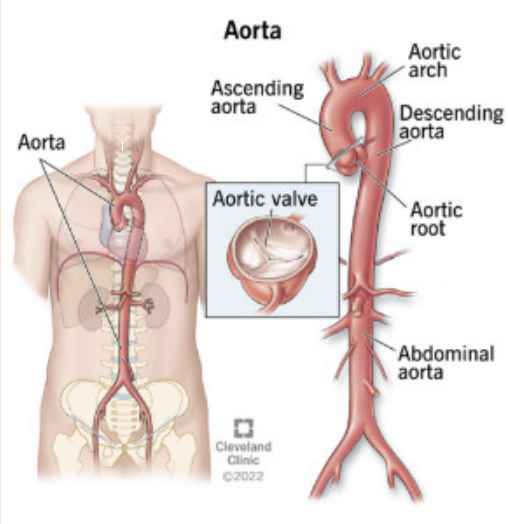
If you have been diagnosed with Marfan and Loeys-Dietz syndromes, you are probably familiar with your Cardiologist who prescribes medication and invites you for regular scans and reviews to ensure that your heart and aorta remain healthy.

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, 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.

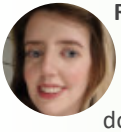
This diagram of the aorta can help you clarify the location of any aortic dilatation or aneurysm you may have and visualise the area of the aorta that your surgeon is planning to treat.

There are many different types of heart surgery that you may undergo as an individual with MFS/LDS, 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

Patient Perspective



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/LDS she has had regular interactions with medical professionalsthroughout 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.

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 prior to your surgery. You can request a dietician referral if you are struggling with this..

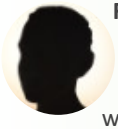
Alcohol – do not drink alcohol to excess. In the UK the recommendation is for both men and women not to drink more than 14 units of alcohol per week. Ideally, you should have several alcohol-free days each week.

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.

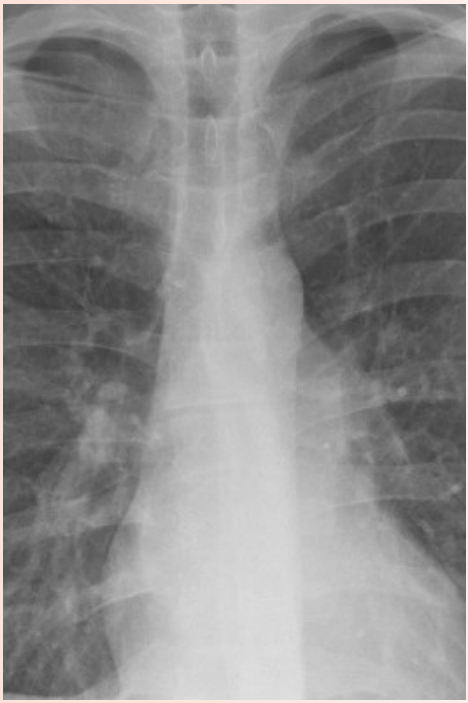
Deep breathing exercises – it can be helpful to start practising taking deep breaths prior to your operation as this is very important once you have had surgery. Take deep breaths whilst sitting upright, hold the breath for a count of three before breathing out, and repeat three or four times during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

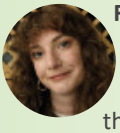
Do you have someone who can collect you from hospital?

Do you have someone living with you?

Can someone stay with you after your operation?

Can you stay with a friend/relative?

Patient Perspective



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 anxietyprovoking 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.

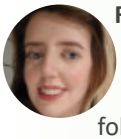
Pre-assessment visit

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. Pregnancy and heart surgery is often a concern for young women and you may have additional questions. Please refer to our Pregnancy Guide for further detailed information: **https://hubble-live-assets.s3.eu-west-1.amazonaws.com/marfantrust/file_asset/file/2842/Pregnancy_Leaflet_2026.pdf**

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.

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.

Back on the ward

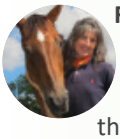
Getting up and about as quickly as possible after your operation is important. This helps to prevent complications such as chest infections, constipation and pressure sores.

- 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.

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.

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

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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.**

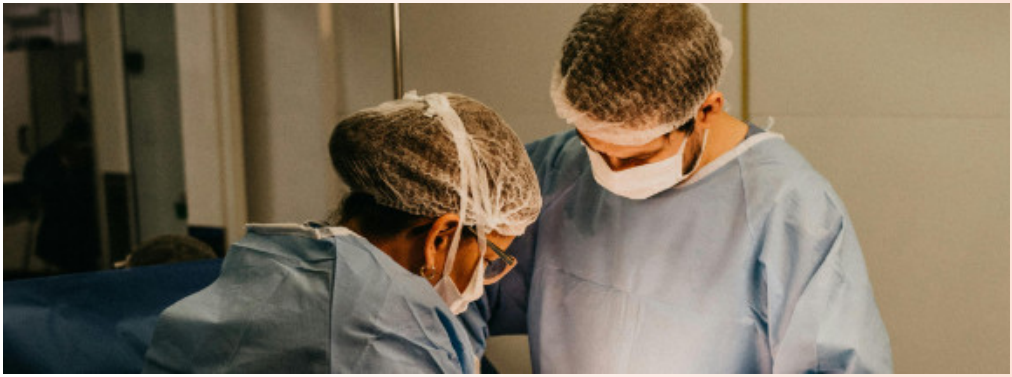
It is normal for your appetite to be reduced after an operation and your sense of taste or smell might be affected for a while. However, it is important to start eating again as your body needs nutrients to heal following surgery. You will be given regular meals and snacks in hospital and **need to keep 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.

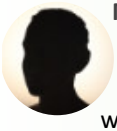
Patient Perspective

Darren underwent aortic root replacement and aortic valve repair (VSRR) 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 ofsurgery 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.

Please visit our Youtube channel:

https://www.youtube.com/@marfantrust3841

Going home

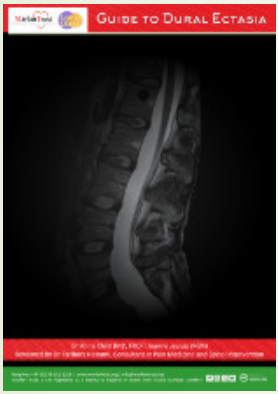
Most people are ready to go home around five to seven days after their operation, but everyone is different, and **the hospital won't send you home until they feel you are fit and well.**

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

Download Our Publications

https://www.marfantrust.org/resources/information-guides-



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 genetic condition, 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.

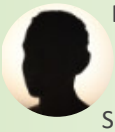
It is important to remember that many patients with Marfan or Loeys-Dietz syndrome will have had surgery to the aorta and this needs long-term follow-up. Lots of hospitals that do aortic surgery will have specialist clinics to monitor patients, so ask your doctor how this will be done and how often you 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.

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, someone with a more manual job will not be able to go back to work so early 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 or Loeys-Dietz 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 or Loeys-Dietz 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 Ration (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.**

Other Investigations

Your aortic valve needs to be protected from infection when anticipating other investigations you may need. Prior to any invasive diagnostic or treatment procedures to another body system (like your lungs, or your gut) you should make your doctor is aware of your increased risk of endocarditis. They can then decide whether antibiotics will be required.

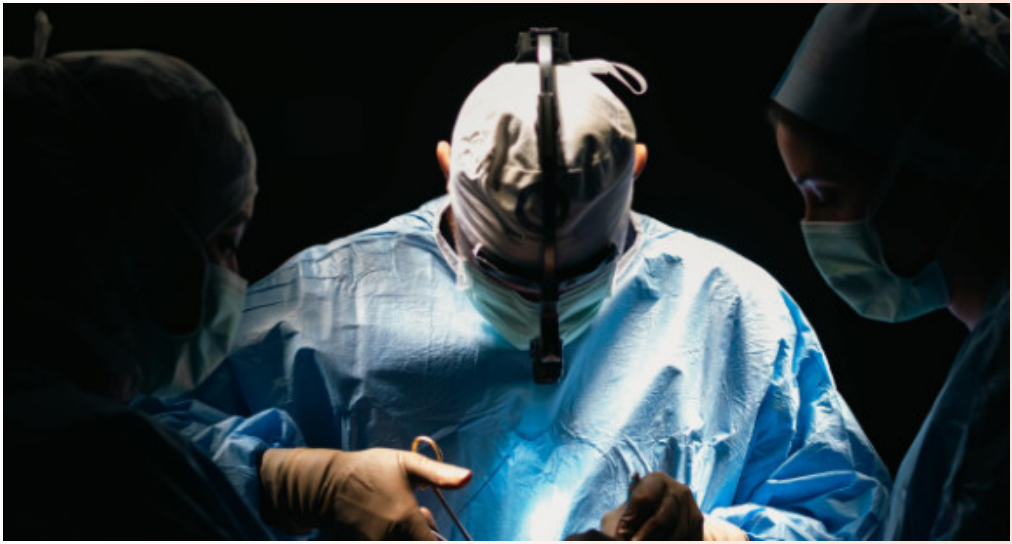
Patient Perspective

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.

Please refer to DVLA guidelines for up to date guidance. **https://www.gov.uk/guidance/cardiovascular-disorders-assessing-fitness-to-drive#marfan-syndrome-and-other-inherited-aortopathies** If you are in any doubt please speak to your surgeon or cardiologist who will be able to advise and complete the appropriate DVLA forms if necessary.

Symptoms to be aware of

Many individuals with Marfan or Loeys-Dietz 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/informationsupport/publications/heart-conditions/ having-heart-surgery**
- British Heart Foundation – Cardiac Rehabilitation Information **https://www.bhf.org.uk/informationsupport/support/practical-support/cardiacrehabilitation**
- 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-youquit/**
- Drinkaware – alcohol consumption **https://www.drinkaware.co.uk/facts/alcoholic-drinks-and-units/how-muchalcohol-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-todrive#marfan-syndrome-and-other-inherited-aortopathies**

How you can help

The Marfan Trust relies solely on the goodwill of its unstoppable supporters who tirelessly raise funds and awareness, allowing the charity to continue its good work and lift the shadow from this condition.

Marfan Trust – Supporting Research into Inherited Aortic and Related Disorders – Loeys Dietz
CONNECTING THE UNCONNECTED AND STRENGTHENING LIVES

You can help to secure the Marfan Trust's future by becoming a member today for just £3 per month:

www.marfantrust.org/pages/10-membership

JustGiving

Just Giving – **http://bit.ly/3Scj51w**

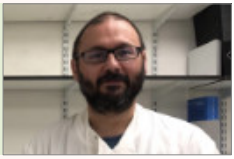
PayPal

PayPal Giving – **https://bit.ly/45NCuwQ**

- BANK: Charities Aid Foundation (CAF) ACCOUNT NAME: Marfan Trust
- SORT CODE: 40-52-40
- ACCOUNT NUMBER: 00017677
- REFERENCE: Your Name (plus campaign name if relevant)

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.

You can also help to fund a piece of equipment in Dr José Aragon-Martin's Sonalee Laboratory. Email **info@marfantrust.org** to find out more.



The Hospital Saturday Fund — Supported by The Hospital Saturday Fund