

# CARDIAC SURGERY

People with Marfan syndrome frequently have problems with their heart and blood vessels. Sometimes, these problems are very serious. The most common complication affects the aorta (the main blood vessel carrying blood from the heart to the rest of the body). Heart valves may be affected as well. Less often, blood vessels other than the aorta are affected. Early and accurate diagnosis is important so that medical issues with the heart and blood vessels can be identified and managed before they become potentially life-threatening emergencies.

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Medical issues related to the heart and blood vessels affect about 9 out of every 10 people diagnosed with Marfan syndrome. The good news is that there are many treatment options, including medication and surgery, as well as adjustments you can make on your own to your physical activity routines. Proper management can help relieve or prevent many of the complications that can interfere with daily life. Medical treatment and ongoing follow-up can usually prevent life-threatening complications.

Most people with Marfan syndrome who have heart problems are helped by elective surgery, which is planned surgery before there is an aortic dissection or other life-threatening problem. The earlier a potential problem is identified and treated, the lower the risk of life-threatening complications.

Surgery is often recommended when the aortic root gets to be a certain size and before it tears (ruptures). Emergency surgery is required if the aortic root tears.

In all cases, it is important that the surgery is performed by doctors who are experienced with operating on patients with Marfan syndrome.

### **What are the common types of cardiac (heart) surgery for people with Marfan syndrome?**

Following are the three most common types of surgery:

- Repair of the ascending aorta. When the aorta reaches a certain size (4.5 or 5.0 cm), surgery to repair the ascending aorta is recommended. All surgery has risks and benefits; however, research shows that repair of the ascending aorta can be very successful when performed by doctors experienced in treating people with Marfan syndrome.
- Aortic or mitral valve repair or replacement. Aortic valve surgery may be recommended when the ascending aorta is replaced. Mitral valve surgery is needed if the mitral valve leaks so much that the heart must pump extra hard. People whose heart valves are replaced with a mechanical valve must take blood thinners (e.g., warfarin or Coumadin®) for the rest of their lives.
- Repair of the descending thoracic or abdominal aorta. This surgery is done when there is a sudden or large change in the size of the descending thoracic or abdominal aorta, or if these parts of the aorta reach a very large size (6.0 cm).

After surgery, people must take blood pressure medication and need tests (such as CT or MRA scans) at least once a year to continue to monitor the size and function of the aorta and heart valves because additional problems could occur and more surgery might be needed. In addition, steps must be taken to prevent endocarditis (inflammation of the heart cavity and valves), which can result when bacteria enters the bloodstream. Dentists and physicians should be alerted to this risk and provide protective medicines before they perform certain dental work or before certain medical procedures (our statement about endocarditis prevention to share with your doctor can be found on our website, [marfan.org](http://marfan.org)).

### **What are the common types of cardiac surgery to repair the aortic root?**

There are three different types of surgery that doctors may use to repair the aortic root. Each has its advantages and disadvantages. We recommend you use a surgeon who has experience with aortic root surgery in patients with Marfan syndrome and talk with him or her about which surgery is best for you.

#### Composite Graft Surgery (Bentall Procedure)

In this surgery, the dilated (enlarged) part of the aorta and aortic valve are removed. They are replaced with a Dacron (synthetic polyester fabric) tube that has a mechanical aortic valve sewn to one end. This surgery has been used for more than 30 years. Results last a long time and, historically, this has been the most common way to repair the aortic root in people with Marfan syndrome.

#### Advantages

- Surgeons have a lot of experience with this type of surgery.
- There are few complications after surgery.
- This repair is very durable; that is, it usually lasts a person's lifetime.

#### Disadvantages

- After surgery, people need to take blood-thinning medication (anticoagulants) for the rest of their lives (and are at risk of bleeding).
- After surgery, people need blood tests to check blood-thinning medication levels every week or two. Tests may be less frequent if the results are the same for a while.
- The mechanical valve is at risk for forming harmful clots and endocarditis (infection of the heart valve).
- There is a clicking sound from the mechanical valve.

#### Valve-Sparing Surgery

In this surgery, the dilated part of the aorta is taken out and replaced with a Dacron tube. The patient's aortic valve is spared (left in place) and sewn into the end of the tube. This surgery is newer than composite graft surgery.

#### Advantages

- People do not need to take blood-thinning medication after surgery.
- There is no risk of forming harmful clots and there is a lower risk for endocarditis (valve infection) than with a mechanical valve.
- There are fewer concerns if a woman becomes pregnant after having this surgery.

#### Disadvantages

- This surgery is a newer procedure and doctors do not yet know how long the repair will last and how long the spared valve will work properly.
- A person may need more surgery if the aortic valve fails (e.g., if it begins to leak severely).
- This type of surgery may not be available in all parts of the country. It needs to be done by a surgeon who has had training and experience performing it in patients with Marfan syndrome.
- This surgery is not an option for everyone. Whether or not a person is a candidate depends on aorta size, whether or not there is damage to the aortic valve, and the amount of leaking from the aortic valve.

#### Bioprosthetic (Tissue) Valve Surgery

In this type of surgery, the aortic valve is replaced with a bioprosthetic (tissue) valve that has been specially prepared.

#### Advantages

- People do not need to take blood-thinning medication.
- Surgeons have a lot of experience with this type of surgery.
- There are several kinds of tissue valves that surgeons can use.
- There is no clicking sound from the tissue valve.

#### Disadvantages

- This type of valve does not last forever, and another operation may be needed in the future.

#### Endovascular Stent-Grafting of the Aorta

Data on stent-grafts in patients with Marfan syndrome and other related disorders is very limited; therefore, there is insufficient information available to guide decisions regarding its safety and efficacy. As a result, stent graft repair is not recommended for people with Marfan syndrome and related disorders.

#### **Do you have questions? Would you like more information?**

- Call our help center, 800-862-7326, ext. 126 to speak with a nurse who can answer your questions and send you additional information.
- Visit our website at [marfan.org](http://marfan.org). You can print information that interests you and ask questions online.

