THE TEETH IN MARFAN SYNDROME

Many people with Marfan syndrome and some related disorders have narrow jaws and high-arched palates, which can create dental and orthodontic problems. In addition, people with mitral valve prolapse and artificial heart valves are at risk for endocarditis (infection of the heart and heart valves) when they have dental work, and should follow recommendations for endocarditis prevention (prophylaxis).

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What are the common types of dental problems in people with Marfan syndrome?

Dental problems that can develop include:

Overcrowded teeth
In Marfan syndrome, the major bone in the upper jaw (the maxilla) is often quite narrow, which can lead to considerable crowding of the teeth.

Posterior crossbite
The narrow high shape of the palate may also cause posterior crossbite—when the upper teeth align inside the lower teeth when you bite down.

Malocclusion
Having a long, narrow face is characteristic of Marfan syndrome. Sometimes this interferes with "facial harmony" and the proper relationship between the upper and lower jaw and teeth (malocclusion).

Tempromandibular joint syndrome

The tempromandibular joint (TMJ) is the hinge that links the jaw to the skull. A misshapen joint or laxity of the ligaments that hold this joint in place can cause a variety of problems that are known as "TMJ syndrome." In all likelihood, people with Marfan syndrome are more prone to TMJ problems, but this has not been studied rigorously.

With TMJ syndrome, you may experience locking of the jaw when it is opened widely, pain when chewing, "popping" or clicking of one or both joints when the mouth is opened, and a persistent ache that can become a headache. A prosthodontist (a dental specialist who replaces missing teeth and restores natural teeth) should be consulted for tempromandibular joint syndrome and jaw problems.

Endocarditis

Endocarditis is the inflammation of the lining of the heart cavity and valves. People with mitral valve prolapse or an artificial heart valve (both are likely in Marfan syndrome and some related disorders) can develop endocarditis during dental procedures and other medical situations where there is an increased likelihood that bacteria can enter the blood stream.

> **Precautions must be taken prior to any procedure that may introduce bacteria into the bloodstream, including routine dental work.**

Many dental procedures go below the gum line and provide an opportunity for bacteria to enter the bloodstream. You should advise your dentist of any heart problems so that your dentist can consult with your cardiologist about the need for antibiotics prior to beginning the dental work.

How are the common types of dental problems treated in people with Marfan syndrome?

The growth and development of the human face is a complex, continuous process that varies from person to person. For children with Marfan syndrome—as with other children—The facial characteristics tend to become more noticeable as the child grows.

The challenge presented by the upper jaw and narrow palate in people with Marfan syndrome is to make the teeth fit and ensure that the upper jaw has the correct relationship with the lower jaw. This is always a challenge for orthodontic treatment, whether or not a person has Marfan syndrome or a related disorder.

Dentists and orthodontists who treat people with Marfan syndrome agree that, for the most part, treatment of orthodontic problems is the same whether or not Marfan syndrome is present. Stability (making sure that the teeth don't move) and retention (maintaining the results of orthodontic treatment) is an issue for everyone and is monitored closely by orthodontists. Treatment for people with and without a connective tissue disorder is truly individualized.

Orthodontic care

There is limited research regarding specific management of the orthodontic problems commonly seen in people with Marfan syndrome, but seeking orthodontic care is an important part of Marfan syndrome management, particularly in children.

According to the American Association of Orthodontists, children should see an orthodontist by the age of 7. This is particularly true for children with Marfan syndrome. Many treatment options

are possible for a growing child. However, as a patient becomes a teenager and an adult, treatment options become more limited.

When a child is 7 or 8, it is possible to recognize a narrow upper jaw, which is a common characteristic in children with Marfan syndrome. A narrow upper jaw causes the upper teeth on the side of the mouth to be set inside the lower teeth, creating a posterior crossbite.

In a young child, it's usually possible to treat posterior crossbite by widening the upper arch with an orthodontic expander. This is because the suture (where the bones of the palate come together) is not yet fused. As a child gets older, these areas fuse and become less flexible. Typically, when a child becomes a teenager, simple orthodontic expansion is not possible without surgery to make the bones flexible again.

However, due to the nature of Marfan syndrome and the continued growth that people with the disorder experience, the time frame for correcting the posterior crossbite with an orthodontic expander varies, and can extend beyond the pre-teen years. If expansion is done early to take advantage of the flexible palate, it is often beneficial to follow this treatment with full braces. If braces are not started right away, there are other options such as a transpalatal arch to hold the space until braces are appropriate.

If a posterior crossbite cannot be corrected with an orthodontic expander (usually because a person is past the growth period), a surgery can assist in the widening of the upper arch.

Any surgical procedure introduces certain risks to people with Marfan syndrome who already have heart-related complications. The patient's heart doctor (cardiologist) should be consulted to evaluate the risk-benefit for this elective (optional) surgical procedure. If the cardiologist and the patient decide surgery is not worth the potential risks, the posterior crossbite cannot be treated. The orthodontist will separately—and nonsurgically—address other orthodontic issues, such as crowding, an overbite, or an underbite.

It is important to consult with your orthodontist to determine the timing that is best for your situation and, if there is any question, get a second opinion from another orthodontist who has treated people with Marfan syndrome.

Endocarditis

Endocarditis is a serious complication for anyone, but particularly for a patient who has had surgical reconstruction of the aorta with placement of an artificial valve, as many people with Marfan syndrome have. It is almost incurable by medicine alone, and nearly always requires surgery to remove the artificial valve and dacron graft. Not only is the operation itself riskier than the original procedure, but also there remains a substantial chance that not all of the infected tissue will be removed and that endocarditis will recur.

To prevent endocarditis, precautions must be taken prior to any procedure that may introduce bacteria into the bloodstream. This includes routine dental work. Tell your dentist that you have Marfan syndrome and about any related heart issues. Then your dentist should consult with your cardiologist about the need for antibiotics prior to beginning the dental work (endocarditis prophylaxis). For more information about endocarditis prophylaxis, please see our statement about Endocarditis Prophylaxis for People with Marfan Syndrome.

Do I need to see a specially-trained dentist or orthodontist?

Children and young teens with Marfan syndrome should first visit a pediatric dentist, who is trained to treat a variety of children, including those with different syndromes. Most general dentists are comfortable treating people with health concerns, but if they are not, they will usually refer you to the nearest university or medical center. If you have any concerns about seeing a general dentist, you may visit a hospital-based dentist, who is accustomed to treating patients with special needs. For orthodontic treatment, you can see a private practice orthodontist because they are also trained to treat people with different conditions—both children and adults.

In many cities there are craniofacial teams, which consist of an orthodontist, pediatric dentist, oral surgeon, speech pathologist, audiologist, and plastic surgeon. If the local orthodontist feels that a case is too difficult to be treated in his/her practice, a referral may be made to the nearest cranio-facial team.

In any case, you should advise your dentist or orthodontist about your condition and explain any cardiac problems that exist. It is especially important that your dentist and orthodontist know about any mitral valve abnormality, aortic regurgitation, artificial heart valves, and heart surgeries. In most cases, they will also request a letter from your treating cardiologist regarding the status of your heart.

What can you do on a day-to-day basis?

There are several things that you can do on a day-to-day basis to care for your teeth when you have Marfan syndrome or a related disorder.

> **First and foremost, daily dental care—brushing and flossing—and regular visits to the dentist are a must!**

Because of the risk of endocarditis, if you have Marfan syndrome you should aim to keep your teeth as healthy as possible before any problems arise to minimize the amount of dental work you need. Gum infections (periodontal disease) provide a breeding ground for harmful bacteria, so keeping gums healthy by practicing good oral hygiene is essential.

You should also:

- Consult a cardiologist about the need for antibiotics before dental procedures.
- Consider if dental or orthodontic work is needed to improve function (eating or speaking) or for cosmetics (to improve appearance). It is important to balance the treatment of dental issues against risks to the heart and cardiovascular system.

Children with Marfan syndrome should see an orthodontist by the age of 7.

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. You can print information that interests you and ask questions online.