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

Marfan Trust Guide to Dental Care

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What is Marfan Syndrome?

Marfan syndrome (MFS) is a genetic disorder of the body's connective tissue that affects any gender, race or ethnic group. Connective tissue helps provide structure to the body, binding skin to muscle and muscle to bone. It provides the stretchy strength of tendons and ligaments around joints and in blood vessel walls. It also supports the internal organs. The tissue Is made of fine fibres and 'glue'. One fibre is called fibrillin.

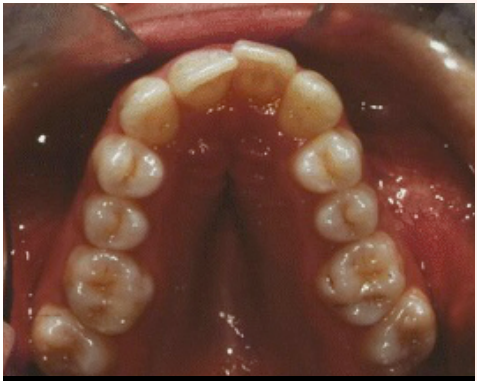
In MFS, a change in the fibrillin-producing gene, FBN1, means that this protein is deficient in connective tissue throughout the body, creating an unusual stretchiness and weakness of tissues. This has far-reaching implications and can affect the eyes, lungs, gut, nervous system, skeleton and, most dangerously, 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.

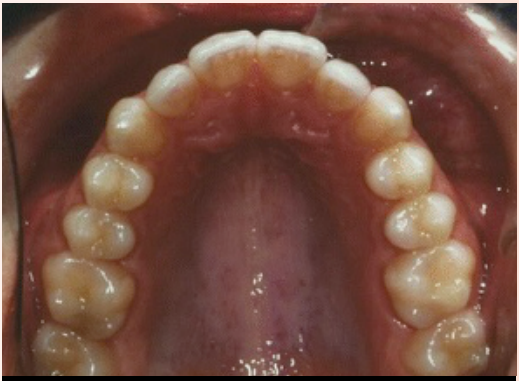
Loeys-Dietz syndrome (LDS) is a connective tissue disorder which overlaps with Marfan syndrome. The dental manifestations are very similar to those in MFS and this guide is therefore relevant to people with LDS. One of the findings that might distinguish between LDS and MFS is that of a bifid (or split) uvula (this is the small fleshy tissue that hangs at the back of the throat).

Lucy has Marfan syndrome

Dental Aspects



Marfan patient showing a high palate and narrow arch with missing teeth



Non Marfan patient for comparison

Some of the craniofacial features seen in MFS can contribute to dental issues that people with the condition can experience, these include:

- High-arched palate
- Temporomandibular (jaw) joint hypermobility
- Growth differences in the shape of the upper and lower jaw bones which can cause issues like crossbite and a greater need for orthodontics (braces)
- Dolichocephaly (having a long, narrow skull)

These features can lead to dental issues:

- Overcrowded teeth and narrow palate
- Temporomandibular joint disorder – this can include pain, locking of the jaw, clicking of the jaw or headaches and jaw tension
- Malocclusion (different relationship between the upper and lower jaws that can affect appearance and the bite)
- Crossbite – the upper teeth align inside the lower teeth

(Cervino et al, 2020)

Developmental abnormalities may also be evident, the most common being the presence of supernumerary (extra) teeth. Rare cases of congenital absence (missing teeth), incomplete development, enamel hypoplasia (poor quality of tooth enamel), dentinogenesis imperfecta (abnormal development of dentine) and multiple odontogenic cysts and pulp stones have also been reported. There is a possibility that the fibrillin defect could contribute to slight relapse after orthodontic treatment, and greater periodontal (gum) problems, often caused by inability to clean overcrowded teeth effectively.

Dental Management

Good oral hygiene along with regular routine dental check-ups are essential. Twice daily brushing along with flossing can help prevent tooth decay and gum infections. Overcrowded teeth can make good oral hygiene more challenging, so time and care need to be taken over brushing.

Prevention is better than cure so all patients should aim to eat a healthy, balanced diet with lower sugar intake to minimise the need for intervention such as fillings or extractions.

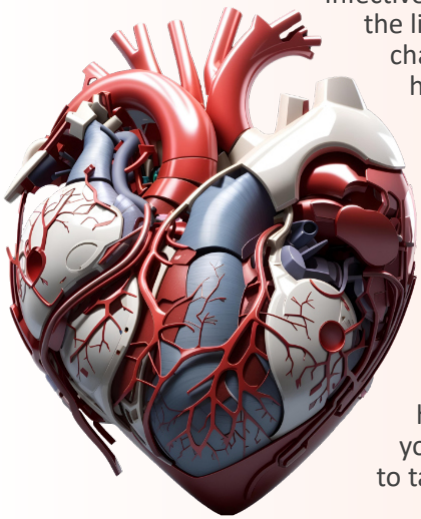
Dental Management

As with most aspects of MFS, the care provided needs to be highly individualised for each person as the condition affects everyone uniquely. The same is true of dental care.

Consideration should be given for different types of local anaesthetic within the dental setting without adrenaline where appropriate.

The features of MFS can become more prominent as a child or young person grows so they need regular review with a dentist or orthodontist to monitor how their jaw and teeth are developing. It is easier to carry out orthodontic interventions in younger people whilst growth is still ongoing, children should have regular reviews and be referred for specialist input early if necessary. Patients can need both orthodontic and possibly oral surgical care. Careful planning from an early age can lead to successful aesthetic results and help prevent more serious dental problems occurring.

Endocarditis

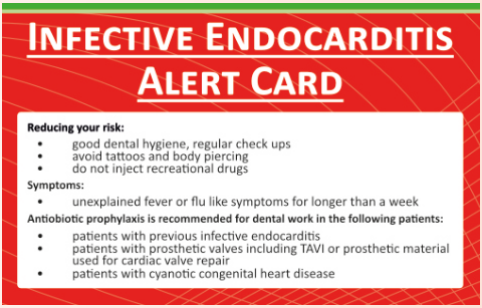


Infective Endocarditis (IE) is a serious condition in which the lining of the heart valves and sometimes heart chambers become inflamed due to infection. This happens when a source of infection (often bacteria) enters the bloodstream and makes its way to the heart. One way in which bacteria can enter the bloodstream is during certain dental procedures. For this reason, preventative antibiotics are needed by people who fall into a high-risk category for IE. A dose is given 30-60 minutes before the dental procedure.

If you fall into a high-risk category, you should have been given an endocarditis warning card by your surgical team and the precautions you need to take should have been explained.

The high-risk categories in both the European (2023) and American guidelines (updated 2021) include:

- People with previous IE
- People with replacement heart valves or those with artificial material used to carry out valve repair
- Those with certain types of congenital heart disease



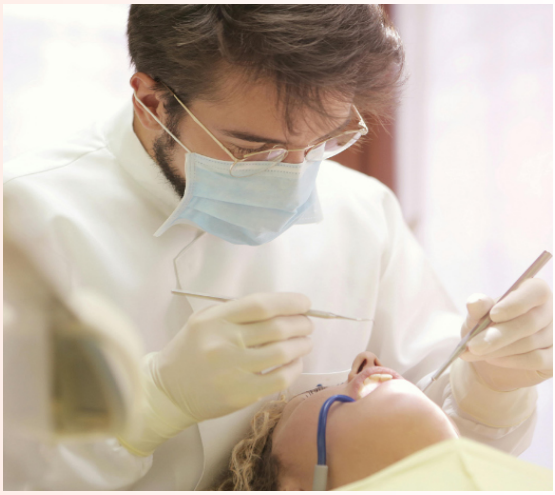
If you are in any doubt about whether you need antibiotic cover for your dental procedure (this includes visits to the hygienist), please contact your dentist who will need to clarify with your cardiologist whether antibiotic prophylaxis is required.

Risk of Bleeding

If you have had surgery to one of your heart valves, you may be taking a blood thinning medication such as warfarin. It is also important to mention this to your dentist prior to any planned procedure as this can increase your risk of bleeding.

General Anaesthesia

Patients with Marfan syndrome are recognised to have a slightly increased morbidity and mortality risk associated with general anaesthesia, due to high narrow palate, reduced neck mobility and narrow trachea causing difficulty with intubation, and increased risk of arrhythmia during anaesthetic. The contributing factors to this are cardiovascular abnormalities, impaired respiratory function, scoliosis, the potential to develop endocarditis and a tendency to spontaneous pneumothorax.



Rarely, difficulty with intubation has been reported due to limited neck extension, a high palate and a narrow trachea. Pre-operative assessment should include a thorough medical examination with a chest x-ray, electrocardiogram and an echocardiogram. Any treatment must be carried out in conjunction with the patient's cardiologist.

Useful References

Cervino et al (2020) Oral health in patients wiht Marfan syndrome, Archives of Oral Biology, 116, 104745

Baddour LM, et al (2021)Infective Endocarditis in Adults: Diagnosis, Antimicrobial Therapy, and Management of Complications: A Scientific Statement for Healthcare Professionals From the American Heart Association, Circulation, Vol 132 (15), **https://doi.org/10.1161/CIR.0000000000000296**

Delgado V, et al (2023) ESC Guidelines for the management of endocarditis: Developed by thetask force on the management of endocarditis of the European Society of Cardiology (ESC) European Heart Journal, Volume 44, Issue 39, 14 October 2023, Pages 3948–4042, **https://doi.org/10.1093/eurheartj/ehad193**

The Marfan Trust

Co-founded in 1988, the Marfan Trust is the sole charity in the United Kingdom dedicated to improving and saving the lives of those with Marfan syndrome. It is estimated that approximately 18,000 people are living with Marfan syndrome in the United Kingdom and we estimate that half remain dangerously undiagnosed.

The Marfan Trust's three main objectives are to:

- provide personalised support and medical guidance through its helpline;
- conduct cutting-edge medical research through its self-funded Sonalee Laboratory, named after a young doctor who tragically died of complications from MFS during her ward round;
- continue to provide educational information and raise awareness of the condition.

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.

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

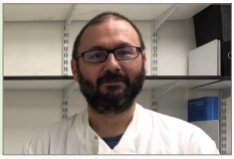
Just Giving – **http://bit.ly/3Scj51w**

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.



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