Slightly increased morbidity and mortality risk associated with general anaesthesia.(6) Preoperative assessments 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.(7)

CONCLUSION

In summary, the dentist must be aware of all the problems associated with treating a patient with Marfan Syndrome.
Prevention of tooth decay, regular check-ups and careful forward planning all reduce the need for more elaborate and potentially dangerous procedures.
The classical marfanoid appearance of the face and mouth can be recognised by a dentist, and could be the first vital step towards diagnosis of the underlying condition. If the diagnosis is suspected, the patient should be referred for echocardiography and genetic counselling through the family practitioner.

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MARFAN SYNDROME SUPPORT GROUP IRL

Founded by a group of people who are directly affected by Marfan syndrome, to provide physical and emotional support to Marfan syndrome sufferers and their families.

The Support Group was also set up to provide information and create awareness for Patients, Relatives, the Medical Profession and the wider community.

TO OBTAIN FURTHER INFORMATION PLEASE FEEL FREE TO CONTACT THE FOLLOWING FOR HELP AND ASSISTANCE

• CORK	Johanna & Petronella	021 4630936
• DUBLIN	Martin & Mary	01 6239563
• KILKENNY	Margaret	056 7767894
• LIMERICK	Daphne	061 377276
• MEATH	Brian & Jo	046 9071232
• MAYO	Caroline & Pat	0979370899
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PATRONS

Dr. John Kenny, Cardiologist

Kate O'Flaherty, Sunday Tribune

Prof. D. Sleeman, Oral & & Maxillofacial Surgeon

Prof. Andrew Green, Geneticist

Prof. Mark Redmond, Cardio-Thoracic Surgeon

Dr. Gary Treacy, Ophthalmologist

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WHAT IS MARFAN SYNDROME

An inherited disorder of connective tissue that affects many organ systems including the skeleton, eyes, heart, lungs and blood vessels.

- *Caused by a mutation in the gene for fibrillin-1 on chromosome 15*
- *Can affect both men and women of any ethnic group*
- *Around 1 in 5000 people have Marfan Syndrome*

SALIENT FEATURES

SKELETAL
Tall, thin physique, with long limbs and fingers, scoliosis, narrow chest and breastbone deformity, joint hyper-mobility and dislocations. Dilation of lumbar dural sac occurs in about 75% of patients.

CARDIOVASCULAR
Dilation of ascending (and sometimes descending) aorta, incompetence of aortic and mitral valves, aneurysm and rupture of aorta.

RESPIRATORY
Phneumothorax, bronciectasis, emphysema and asthma.

OCULAR
Dislocation of lens, myopia and unstable refraction, detachment of retina, squint, glaucoma.

DENTAL
High arched palate, crowding of teeth.

LUNGS
In some cases Pneumothorax (collapse of the lungs) may occur requiring hospital treatment. Recurrent pneumothorax may be treated to prevent further episodes

GENETIC
Males and females are affected with equal frequency. Each child of an affected parent has a 50% chance of inheriting Marfan syndrome.
In about 25% of cases neither parent is affected; however, apparently unaffected parents should be screened carefully, as the severity and pattern of disease are variable even within one family

DIAGNOSIS

Diagnosis is made after careful physical examination and echocardiography, demonstrating classical features in two out of three major systems ; eyes, heart, skeleton, preferably with a family history. Diagnosis can be confirmed within a family by genetic linkage studies. Mutations can be found in the fibrillin-1 gene in 80% of patients, assisting with screening of family members.

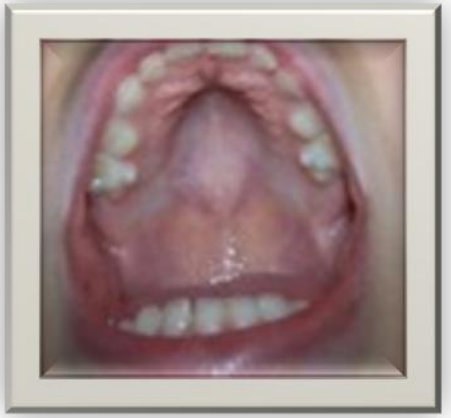
CARDIAC PROBLEMS

The most serious problems occur in the heart and blood vessels. The aorta is usually wider than expected and is more fragile. The dilatation tends to be progressive, leading to aortic regurgitation and dissection. Surgical repair is recommended when the aortic root reaches 5cm, or earlier in cases with a family history of earlier dissection. Beta-blocker therapy can delay dilatation. Mitral valve prolapsed is often also present. Antibiotic prophylaxis is recommended for all dental invasive procedures.

DENTAL ASPECTS

The clinical findings in the oral region display the same great variability in expression as is shown throughout the rest of the body. The most common oral sign is a high arched palate. Collapse of the upper dental arch, and in some cases severe malocclusion, cross bites and open bite, tend to be associated with the high palate.
Other important characteristics include a long, narrow face and skull (dolicocephaly), often with mandibular prognathism, prominent supraorbital ridges, deep-set eyes and frontal bossing. (1,2)
Weakened capsular ligaments and hyper-extensibility of muscle can contribute to dysfunction of habitual dislocations or sublimations of the temporomandibular joint. (1,2,3)
Developmental abnormalities may also be evident, the most common being the presence of supernumerary teeth. Rare cases of congenital absence, incomplete development, crown dysplasia, enamel hypoplasis, dentinogenesis imperfecta and multiple odontogenic and cysts have also been reported.

There is a possibility that the fibrillin defect could contribute to slight relapse after orthodontic treatment, and greater periodontal problems.



DENTAL MANAGEMENT

Good oral hygiene is essential, along with regular routine dental check-ups. Patients may also need 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.(4)
The prevention of bacterial endocarditis is all important because of the high incidence of heart valve involvement and abnormalities of the great vessels in Marfan syndrome. Therefore antibiotic prophylaxis is mandatory for those who have cardiac problems requiring antibiotic cover and one should obviously check with the patient's cardiologist.