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MARFAN SYNDROME

What is Marfan syndrome?

Marfan syndrome is an inherited disorder that affects "connective tissue". Connective tissue supports tendons, ligaments, blood vessel walls, cartilage, heart valves and other tissues. Marfan syndrome causes the affected tissues to be less stiff and supportive as they should be. Marfan syndrome affects many parts of the body, but this discussion focuses on the cardiovascular effects of Marfan syndrome.

Marfan syndrome may be very mild or severe. There is no single test for diagnosing the disorder, but doppler ultrasound and chest X-rays are frequently used in follow-up of individuals with the condition. People who have the condition share some similar traits, including:

- heart problems
- tall, thin stature
- long arms and legs
- slender fingers
- spine curvature
- eye problems

How does Marfan syndrome affect the cardiovascular system?

In Marfan syndrome, the walls of arteries are weakened. The aorta (the large artery that leaves the heart) often becomes enlarged, with a weakening of the inner wall of the aorta. The aortic wall can tear and/or dissect. Marfan syndrome also increases the possibility of developing an aneurysm (a ballooning out of a blood vessel, which can cause clots or rupture).

Marfan syndrome can involve the heart valves. Blood may leak backward (regurgitate) through a valve, which increases the heart's workload. This increased workload can cause the affected area of the heart to enlarge. Mitral valve prolapse is a common but less dangerous condition associated frequently with Marfan syndrome.

How is Marfan syndrome treated?

Some people with mild symptoms might not need specific treatment beyond routine follow-up with their doctor. Medications to lower heart rate and/or blood pressure are sometimes indicated. When heart valves are significantly affected, surgery is sometimes indicated

Marfan syndrome increases the possibility of developing endocarditis, which is an infection of the endocardium (the tissue that lines the heart wall and valves). To prevent this, antibiotics are often prescribed before medical or dental procedures. Ask your doctor about this if you have any questions or think you might need antibiotic therapy for an

upcoming procedure. These procedures include something as minor as getting your teeth cleaned at the dentist.

People with Marfan syndrome might need to modify their physical activity, depending on the severity of their condition. Contact sports that could result in a blow to the chest are usually to be avoided, as well as weight lifting and most team sports. Ask your doctor about your particular situation.

Pregnancy for women with a dilated aorta is not advised, because of the stresses of pregnancy and delivery.