Commentary

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Treatment of Marfan Syndrome

Marathe*

Department of Internal medicine, Ain Shams University, Egypt

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***Corresponding author:** Marathe, Department of Internal medicine, Ain Shams University, Egypt

Tel: +447817502236

rmart@nhs.net

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Introduction

Marfan syndrome is a genetic condition that affects connective tissue, which provides support for the body and organs. Marfan syndrome can damage the blood vessels, heart, eyes, skin, lungs, and the bones of the hips, spine, feet, and rib cage.

The treatment of Marfan syndrome is directed toward the specific symptoms that are apparent in each individual. Treatment may require the coordinated efforts of a team of specialists including geneticists, surgeons, cardiologists, dental specialists, eye specialists (ophthalmologists), orthopedists, and other healthcare professionals.

Individuals with Marfan syndrome are encouraged to avoid competitive and contact sports, heavy lifting and any exercise that increases the strain on the aorta produced by rapid or vigorous beating of the heart or increased blood pressure. Restriction of such activities can slow the rate of the widening of the aorta (aortic dilatation) and decrease the tendency for aortic tear (dissection). In general, moving types of exercises performed in moderation are thought to be good for people with Marfan syndrome. Such exercises, performed regularly, will naturally lower heart rate and blood pressure.

Beta-adrenergic receptor blocking drugs (β-blockers) such as propranalol or atenolol are often used in treating the cardiovascular problems associated with Marfan syndrome. Such drugs help to reduce the strength and frequency of the contractions of the heart. In doing so, they may reduce the strain on the walls of the aorta. Beta-blockers may delay the need for heart surgery. The dosage needs to be adjusted to the individual patient's needs, and therapy should be closely monitored. Some individuals may not be able to tolerate these drugs and others such as those with asthma or depession may not be able to take them (contraindicated).

A second class of blood pressure medication called angiotensin receptor blockers (ARBs) is commonly used in the treatment of cardiovascular problems associated with Marfan syndrome. This includes medications such as losartan or irbesatan. There is experimental evidence that ARBs can help by both lowering blood pressure and by blocking TGF-β activity. In animal models of Marfan syndrome the protective effects of ARBs was superior to that seen with β-blockers. In clinical trials, ARBs have variably been shown to be either better than or as good as β-blockers in suppressing aneurysm growth, but this may not be true for all patients or in all circumstances. In the largest trial performed to date, young patients receiving β-blockers (at high dosing) or ARBs (at standard dosing) had a comparable decline in the deviation of the aortic root size from that expected for age and body size (decreasing aortic root z-score). While both treatments were well tolerated in this study, in general, ARBs are thought to be better tolerated than β-blockers. It is the stated position of the Marfan Foundation that the choice of treatment should be guided by the particular circumstances. A combination of β-blocker and ARB therapy can be considered in circumstances where one or the other type of medication does not achieve an adequate response.

Every person with Marfan syndrome should have at least a yearly echocardiogram to check the size and function of the heart and aorta. Surgical repair of the aorta may eventually become necessary if the aorta has severely widened or developed a tear (dissection). Preventive (prophylactic) surgery is recommended when the diameter of the aorta reaches 5 centimeters in older children or adults, when the rate of widening reaches 1 centimeter a year, or when there is severe or progressive backflow (regurgitation) of blood through the aortic valve. Surgery may also be necessary for leakage of the mitral valve. Replacement of the aortic valve may be performed; however, this surgery requires the lifelong use of medications to prevent blood clots (anticoagulation). In recent years, some physicians have preferred to use valve-sparing surgery (i.e., reimplantation of the natural aortic valve within a Dacron tube used to replace the enlarged segment of the aorta). Studies are underway to assess the durability of valve-sparing procedures, but early data are encouraging.

Surgery to repair or replace the mitral valve in individuals who experience severe mitral valve regurgitation may become necessary. Cardiovascular problems related to Marfan syndrome increase affected individuals' susceptibility to repeated bacterial infections such as infections of the heart valves (bacterial endocarditis). Leaking heart valves are more prone to infection with bacteria. While it had been common practice to treat patients with leaking valves with antibiotics before dental work or other procedures expected to contaminate the blood stream with bacteria, the American Heart Association recently withdrew this recommendation for most people. Given the predisposition of people with Marfan syndrome and other connective tissue disorders to progressive leakage through multiple heart valves, many physicians who routinely care for such

individuals continue to recommend that antibiotics be used before dental work or other procedures expected to introduce bacteria into the bloodstream.

Skeletal abnormalities such as scoliosis and deformity of the chest may represent serious problems for people with Marfan syndrome. Braces may be tried to correct skeletal curving (scoliosis) in some cases, but can be ineffective. Individuals with curvature of the spine of more than 10 degrees should be followed by an orthopedist. Surgical stabilization of the spine may be needed if the curvature is severe or progressive. A sunken chest (pectus excavatum) may be surgically corrected for cosmetic reason or, in very rare severe cases, to avoid medical complications.

The eyes require careful attention (e.g., yearly ophthalmologic exams) from early childhood. Failure to detect any of the several abnormalities that can affect the eyes may result in poor vision and other visual impairment. Increased risk of retinal detachment does demand special attention. The eyes should receive special protection from injury during work or sports. Sports that may involve trauma to the head, such as football, boxing, and diving, should be avoided. Displacement of the lenses may be treated with eyeglasses or contact lenses. Some individuals such as those with a completely loose lens or with a displaced lens that disrupts vision may require surgical intervention. A detached retina can sometimes be corrected, especially if detected early.

Genetic counseling may be of benefit for affected individuals and their families. Other treatment is symptomatic and supportive.

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None

Conflict of Interest

The author declared that there is no conflict of interest.