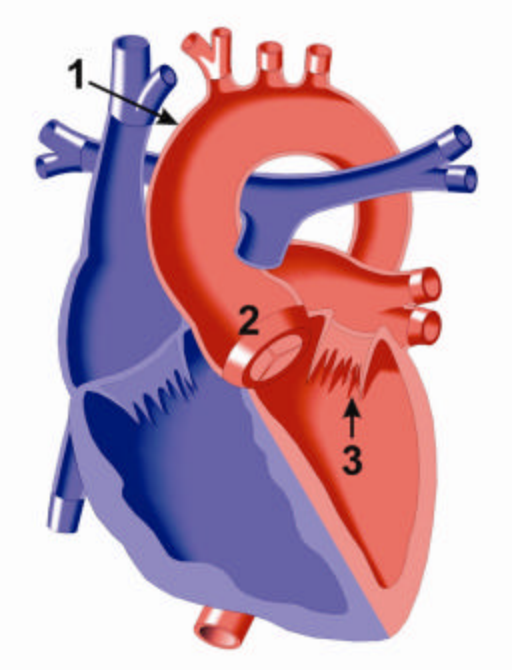
Marfan's Syndrome

This inherited congenital heart defect involves the enlargement of the aorta - the large vessel that carries blood from the heart to the body's tissues. This enlargement occurs near the aortic valve where the aorta leaves the heart and may also affect the part of the aorta that carries blood to the body (the ascending aorta).



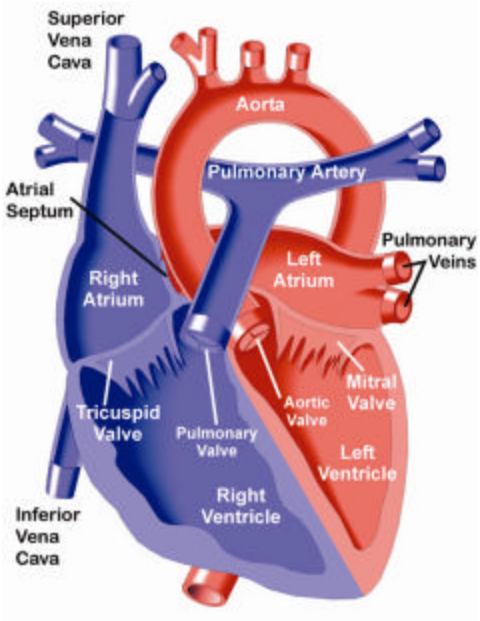
There may also be abnormalities in the structure of the aortic valve and/or the mitral valve, which connects the left atrium and left ventricle.

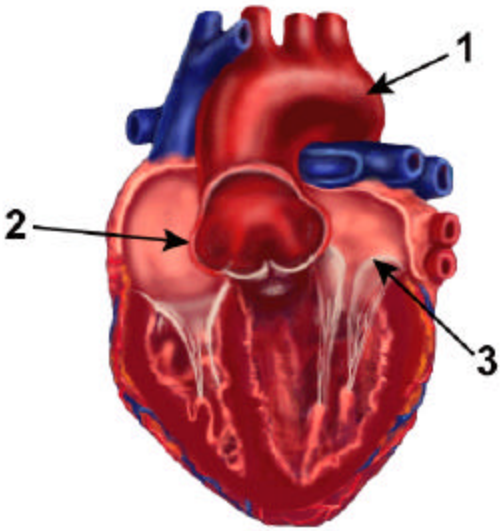
Marfan Syndrome (also known as arachnodactyly) is named for Dr. Antonin Marfan, who first described this condition in 1896. It is caused by a genetic defect involving the body's connective tissue and occurs in approximately 1 of every 10,000 people.

There is progressive enlargement of the aorta, caused by the high blood pressure in this vessel as its wall becomes thin and weak. Enlargement of the aorta can lead to rupture, sometimes resulting in sudden death.

If the structure of the aortic or mitral valves is abnormal, there may be the leakage of blood at these points, resulting in fatigue, shortness of breath, and an irregular pulse.

People with Marfan are often very tall and double-jointed, with proportionally long arms, legs, and fingers. They generally have weak connective tissues (such as tendons and ligaments) in various parts of the body, including the skeleton, eyes, heart, and other organs. Besides the difficulties in the heart already described, this may result in curvature of the spine and dislocation of the eye lenses. The severity of the symptoms of Marfan Syndrome is variable.





Top and Above:
1. Dilated aorta
2. Dilated sinuses of Valsalva
3. Mitral valve prolapse

Left: Normal Heart