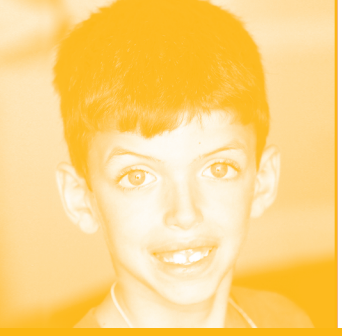

Cardiovascular Features:

Common Types of Heart and Blood Vessel Problems in People with Marfan Syndrome

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Cardiovascular (heart and blood vessel) problems are common in people with Marfan syndrome. In fact, cardiovascular problems affect about 9 out of every 10 people diagnosed with Marfan syndrome.

The most common of these problems affects the aorta (the main blood vessel that carries blood away from the heart). Heart valves can also have problems. Less often, people have problems in blood vessels other than the aorta.

Here is a diagram that shows the main sections of the heart and aorta. As you can see, the aorta has four segments: 1) aortic root and ascending aorta, 2) aortic arch, 3) descending thoracic aorta, and 4) abdominal aorta.

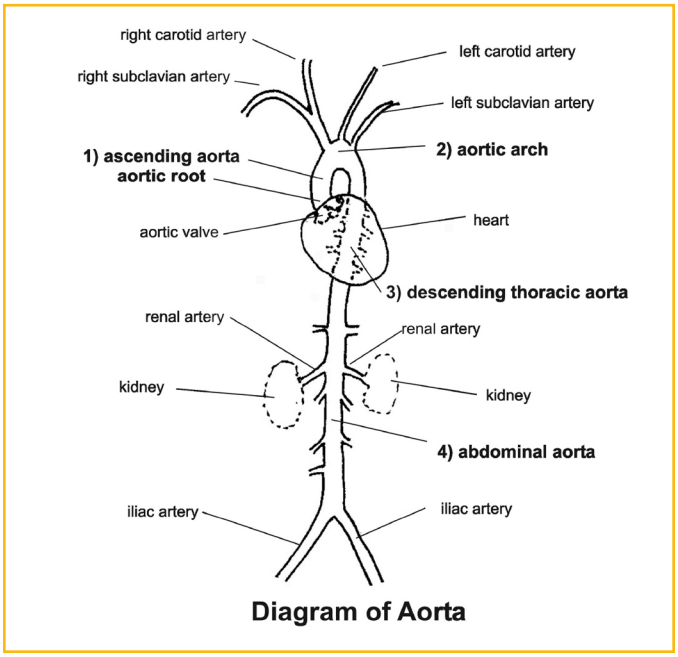

Diagram of Aorta

Here are some facts about common types of heart and blood vessel problems in people with Marfan syndrome:

Aortic Dilation and Aortic Aneurysms

These are very serious cardiovascular problems because a significantly enlarged aorta is at risk for dissection (tear) or rupture. Aortic dilation (enlarged aorta) and aortic aneurysms (bulging sides of the aorta) can occur along any segment of the aorta. For most people with Marfan syndrome, the problem starts in the aortic root (aortic segment closest to the heart.)

Doctors use a person's age, height, and weight to determine whether the aorta is enlarged. For this reason, people with Marfan syndrome should talk with their doctors about what size aorta is within normal limits for them.

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Aortic Dissection

This happens when there is a tear between layers of the aorta. Most people know when this happens because of severe pain in the center of their chest, abdomen (stomach), or back. The pain may be "severe", "sharp", "tearing", or "ripping" and may travel from the chest to the back and/or abdomen. Sometimes, the pain is less severe, but a person still has a feeling that "something is very wrong." If a dissection is suspected, a person needs to go to a hospital emergency room right away. There are two types of aortic dissection:

- Dissection of the ascending aorta. This is the most common dissection in Marfan syndrome. It is life-threatening (people can die if not treated right away). If this kind of dissection happens, people need immediate surgery.
- Descending aortic dissection. This can often be managed just with medication and monitoring (watching). People only need surgery if they have serious complications. These include loss of blood flow to vital organs or an aorta that is severely dilated.

Mitral Valve Prolapse

This is a "billowing" (motion) of the mitral valve when the heart contracts. Symptoms can include irregular or rapid heartbeats and shortness of breath. Some people also have mitral valve regurgitation (leaking of the mitral valve). A small amount of leaking is often not a problem, but a person may need surgery if the mitral valve leaks a lot.

Aortic Regurgitation

This is when the aortic valve does not fully close and blood leaks back into the heart. The only symptoms a person may have are forceful heartbeats and shortness of breath during light activity. Aortic regurgitation often happens because of aortic dilation (when the aorta is so enlarged that the valves cannot fully come together).

Cardiovascular problems in people with Marfan syndrome can be very serious. The good news is that there are many ways to help. This includes surgery and medication. New research studies show that many people with Marfan syndrome may live normal life spans. New research is also finding more helpful medications.

Ways to Learn More

- Contact the Marfan Syndrome Support Group Ireland at info@marfan.ie.
- Talk to your doctor. Sometimes it helps to use information like this fact sheet when you speak with the doctor.
- Visit the Marfan Syndrome Support Group website at www.marfan.ie.