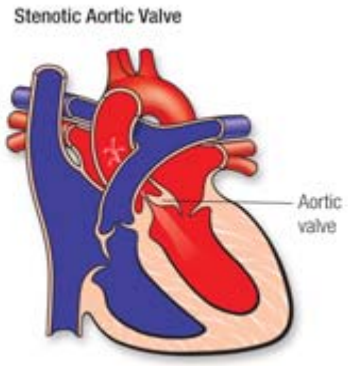
Aortic Valve Stenosis and Regurgitation

(Note: before reading the specific defect information and the image associated with it, it will be helpful to review normal heart function.)



What is it?

The aortic valve is the final door blood goes through as it exits the heart. It opens to let blood out from the left ventricle (the heart's main pump) into the aorta (the main artery bringing blood throughout the body). Obstruction is called stenosis and leakage of the valve as it closes after each heartbeat is called regurgitation or "insufficiency". These problems can occur alone or together.

What causes it?

A normal aortic valve has three leaflets or cusps (tricuspid). About 1 percent of the population is born with a valve that only has two leaflets (bicuspid) and narrows or leaks over time. Another version is a single leaflet valve, which occurs rarely. Genetic factors have been shown to play a role.

How does it affect the heart?

Some infants are born with severely narrowed aortic valves need early treatment. However, most bicuspid aortic valves work normally for a long time — sometimes a lifetime. In other patients, the valve can become thick and narrowed/obstructed (stenotic) or curled at the edges and leaky (regurgitant or insufficient). When the valve is obstructed the left ventricle pumps at a higher pressure than normal to push the blood through the narrow opening. In response, the heart muscle gets thicker. When the valve primarily leaks, the ventricle has to pump more blood and the ventricle enlarges.

How does it affect me?

With mild obstruction, patients usually have no symptoms. The problem is detected by the presence of a clicking sound in the heart and a murmur. When the valve opening narrows to about one-fourth its original size, symptoms are common. The most common symptom of an obstructed or leaky aortic valve is shortness of breath with exertion. This usually develops gradually over time, and some patients will just feel "out of shape." Chest pain, lightheadedness or fainting may also occur. Recurrent fevers may indicate the valve is infected.

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What if aortic stenosis or regurgitation is still present? Should it be repaired in adulthood?

When the aortic valve becomes excessively obstructed (stenotic) or leaky (regurgitation or insufficiency), the valve must be repaired or replaced.

Repair of the obstructed or narrowed valve can be done 1) in the catheterization lab (referred to as interventional or therapeutic catheterization) using a balloon to force the leaflets open; or 2) in the operating room by open-heart surgery. Some valve leakage is likely to develop after either approach. Some patients will have had one or both of these procedures as an infant or child.

Replacing the valve requires open-heart surgery. In most adults, when the valve is no longer working properly, it's best to replace the valve rather than repair it. Deciding when to perform aortic valve surgery and the type of valve to insert are complicated decisions for your doctor. Your aortic valve can be surgically replaced with any of these:

1. A mechanical valve made of metal, which requires you to take blood thinners but is very durable;
2. A valve made from biological tissue, which requires no blood thinners but may not last as long and may need to be replaced later;
3. A homograft or valve from a donated human heart and preserved in special solutions may be use. Homograft valves require no blood thinners but may not last as long (sometimes this is a good option when a portion of the aorta is also being replaced);
4. Your own pulmonary valve (known as the "Ross procedure"), which is then replaced by a preserved donor valve. This requires no blood thinners, and hopefully a more durable aortic valve, although the new pulmonary valve will likely need to be replaced in the future with further surgery.

Each option has advantages and disadvantages. You should discuss them at length with your cardiologist and cardiac surgeon to find the best option for your situation.
Sometimes narrowing is present just below the aortic valve, with or without affecting the valve itself. Then, a more complex surgery, often called a "Konno procedure," may be needed. It will enlarge the part of the left ventricle that leads to the aortic valve. The problem may recur years later, requiring further surgery.

Ongoing Care:

Medical

Everyone with an aortic valve defect needs routine follow-up, whether before or after surgery. Progression occurs over time. Even with the best surgery, the patient is never "cured." The severity of your valve problem will dictate how often you'll need to visit the doctor and how often echocardiograms are needed. Medicines might be useful to lower blood pressure or maintain the health of the left ventricle. You should also consult a cardiologist experienced in caring for adults

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with congenital heart disease if you are undergoing any type of non-heart surgery or invasive procedure.

Activity Restrictions

If you have a severely obstructed valve, vigorous exercise is not a good idea (for more information see the Physical Activity and Exercise section.) Your cardiologist may tell you to limit your activity if this is the case. Ask your cardiologist about your exercise limits.

Endocarditis Prevention

People with even mildly abnormal aortic valves are at risk for bacterial endocarditis. That's why it's important for you to keep your mouth clean and healthy with regular dental check ups. Getting antibiotics before dental procedures isn't proven to be beneficial and so isn't universally recommended any more. But if you have a prosthetic valve, you'll need to take antibiotics before dental work. See the section on Endocarditis for more information.

Problems You May Have

Most patients feel well. Aortic stenosis (obstruction) and insufficiency (leak) usually cause symptoms only when these defects are severe. Symptoms include shortness of breath, exercise intolerance, dizziness, chest pain or occasionally abnormal heart rhythms. Patients may also develop an enlarged aorta over time, which may eventually require surgery. There are usually no symptoms associated with this, which can only be detected with imaging.

Pregnancy

The risk from pregnancy depends on how severely the valve is obstructed or how much it's leaking. If you have mild or moderate stenosis and your left heart muscle (ventricle) is functioning normally, you can have a safe pregnancy, but you need medical supervision throughout the pregnancy. Sometimes balloon valvuloplasty can be done to relieve symptoms if they occur during pregnancy but only when symptoms can't be controlled by medication and bed rest.

If your stenosis is severe and you have symptoms, avoid conception until you've had your heart valve repaired or replaced. If you're considering pregnancy and you have aortic valve stenosis, you should meet with a multidisciplinary medical team that can give you more information about the risk of pregnancy to you and your baby.

Pregnancy in aortic regurgitation is better tolerated, but if the regurgitation has weakened the heart muscle and signs of heart failure are present before pregnancy, the risk posed by pregnancy is higher.

In patients who have had their heart valve replaced with a metal (mechanical) heart valve, they may be taking warfarin (Coumadin) which can cause risk to the fetus and alternative means of blood thinning may be required. In aortic insufficiency, women may be taking medicines such as ACE inhibitors such as lisinopril (Zestril) or enalapril (Vasotec). These drugs are dangerous to the developing fetus (see the section on Pregnancy) and need to be changed before conception. It's best to talk with your doctor before you plan to become pregnant.

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Will you need more surgery?

If you already had an operation on your aortic valve, chances are high that you may need another. This may be to replace a valve or an enlarging aorta. No surgical fix is ever perfect, so regular follow-up with a doctor who is informed about this particular problem is recommended.

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