

What is a bicuspid aortic valve (BAV)?

The aortic valve is the valve in the heart that allows blood to flow from the heart to the aorta, which is the blood vessel that brings the oxygen-rich blood to the body. It usually has three leaflets. In a bicuspid aortic valve, there are only two.

How common is BAV?

BAV is one of the most common heart defects. It occurs in 1% - 2% of the population. It is more common in males than females.

What causes BAV?

The exact cause is not known. It can occur spontaneously or run in families. Not every family member will have it. If you have BAV, your parents, siblings, and children should consider being screened. Medical experts now suggest that BAV may be associated with a connective tissue disorder similar to that found in people with Marfan Syndrome.

What other heart problems can occur with BAV?

BAV often occurs alone. In some people, it is associated with other more complex heart defects. These include coarctation of the aorta (CoA), ventricular septal defect (VSD), subaortic stenosis, Shone's syndrome, and hypoplastic left heart syndrome.

Why do we worry about BAV?

First, the BAV may not close completely and thus become leaky (regurgitation). The BAV may not open completely, and thus become narrow (stenosis).

These problems can occur either earlier or later in life. When symptoms occur between birth and 30 years of

age, the aortic valve is likely inherently abnormal. If they show up between 50 and 60 years of age, the valve probably has become "calcific." While calcific aortic valve disease is common in older adults, a person with BAV can develop it earlier. These issues can make the left ventricle (the primary pumping chamber) become larger and/or get thicker because it has to do more work.

Other signs and symptoms include:

- Heart murmur
- Shortness of breath with exercise
- Dizziness with exercise
- Fainting
- Chest pain or tightness
- Endocarditis

Patients may first notice symptoms during exercise. Later on, they may become short of breath with little or no activity. Some people have trouble sleeping when flat or even wake up short of breath.

Why do we worry about the aorta?

Patients with BAV may have an enlargement or an aneurysm in the ascending aorta (aortic root). This puts them at increased risk for aortic tearing (dissection) or aortic rupture.

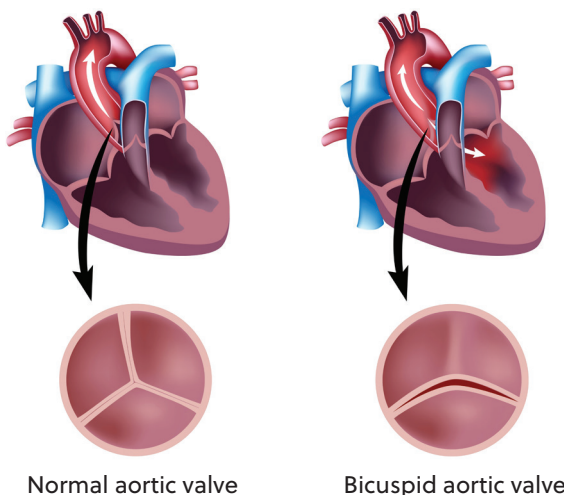
There are two reasons for aortic problems. The first is because of something inherently abnormal in the wall of the aorta. When the aortic wall forms, something genetic causes abnormal features of the vessel. In turn, it becomes weaker and more prone to stretch. External forces also affect the vessel wall. These include the way that blood flows out of the aortic valve and the pressure it puts on the vessel wall. This leads to progressive aortic enlargement.

A normal aorta is 2.5 cm - 3.5 cm in diameter. If it is bigger than 3.5 cm - 4.0 cm, doctors begin to get concerned. The rate of growth varies. Larger aortas grow faster. If you have an aortic aneurysm, you are at increased risk for aortic dissection if you have any of the following:

1. Rapid growth of the aorta (greater than 0.5 cm/year)
2. History of a repaired coarctation of the aorta
3. Family history of dissection
4. History of smoking

How is BAV diagnosed and monitored?

BAV is diagnosed in a number of ways. These include physical exam, EKG, and imaging methods, including echo, MRI and/or CT scan.



Normal aortic valve

Bicuspid aortic valve

Physical exam can tell your doctor a lot. A murmur might tell him/her if the valve is narrow or leaking.

An EKG can tell your doctor if your left ventricle is enlarged. Neither physical exam nor EKG can tell the doctor anything about the size of the aorta.

The 2014 ACC/AHA Guidelines for BAV recommend that patients with BAV have a transthoracic echo (TTE). This test measures how severe the aortic stenosis or aortic regurgitation is. It will also tell your doctor the size of the aortic root, how big the left ventricle is, and how well it is working. It can also tell if there is an aneurysm in the ascending aorta.

If an echo is not able to measure the size of the aortic root, then a CT or MRI is recommended. Serial imaging is indicated if the aortic root is equal to or greater than 4.0 cm. If it is greater than 4.5 cm, advance imaging is recommended every year. If you have serial imaging, an MRI is preferred since it doesn't subject you to repeated radiation.

If you or a family member have BAV, it is recommended that your children and other first-degree relatives be screened for BAV and for aortic dilation.

What are guidelines for intervention if I have an aneurysm?

Surgical replacement of the aorta is recommended when the aortic root diameter is greater than 5.5 cm. It is suggested when the diameter is greater than 4.5 cm if it is growing rapidly or if there is a family history of dissection. If the aortic diameter is greater than 4.5 cm and the patient is having the valve replaced, surgery for the aneurysm would be reasonable.

If I have a BAV, can I have children?

If you have BAV, you should see an ACHD cardiologist before getting pregnant. If your valve is leaking or narrow, or if your aorta is bigger than normal, an ACHD cardiologist can share your particular risk and guide you through the decision. It's important to take care of any issues before you get pregnant.

If I have a BAV, do my children have a higher risk of having a heart defect?

All adults with congenital heart disease are more likely than the general population to have a child with a CHD. The defect can be the same or different than the parent's defect.

If you or a family member have BAV, it is recommended that your children and other first-degree relatives be screened for BAV and for aortic dilation.

How is BAV treated?

In some cases, people may never know they have BAV. It is not found until autopsy. In the most severe cases, when symptoms are present at birth or in early infancy, surgical repair of the valve must be performed immediately. Other people develop symptoms over the course of time. Some will never need surgery. Others will require surgical treatment to repair or replace the valve and/or part of the aorta as they get older.

If valve replacement surgery is necessary, there are two traditional valve options—mechanical or tissue. Mechanical is more durable. However, blood clots are prone to form on them. Patients must take the blood thinner warfarin for the rest of their lives. Tissue valves do not last as long but are less likely to cause blood clots. They deteriorate over time and will eventually need to be replaced. Aspirin is the only blood thinner required.

Specialized procedures include either repair of the BAV or the Ross procedure. In the Ross procedure, the aortic valve is replaced with the patient's pulmonary valve. The pulmonary valve is then replaced with a homograft or cadaver pulmonary valve.

Your ACHD cardiologist will talk about the benefits and risks of each type of valve option and discuss which valve may be appropriate for you.

Can I exercise?

Some patients with BAV, but not all, have exercise restrictions. If your aorta is enlarged, activities that cause you to strain may not be recommended. These include heavy lifting and competitive athletics. Alternately, aerobic exercises, such as swimming, jogging, and biking, are generally safe for most patients.

It is important to talk to your ACHD cardiologist about what the exercise options that are best for you. He/she can give you an exercise prescription.

What kind of heart care is recommended for adults with repaired BAV/valve replacement?

Lifelong follow-up with an ACHD provider is important for all adults with CHD. This includes those with BAV. It is crucial that you see a heart doctor who is trained in and specializes in ACHD. He/she will decide how often you need to be seen and how often you need imaging. You should discuss the appropriate timing of your visits and tests with your ACHD cardiologist.

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