CONDITION

Bicuspid aortic valve

Learn how a bicuspid aortic valve can affect heart and aorta function, including symptoms, testing, monitoring, treatment, and when to seek urgent care.

WHAT IT IS
Congenital aortic valve with two flaps

BODY SYSTEM
Heart and circulation

MAIN SYMPTOMS
Shortness of breath, chest discomfort, unusual tiredness, dizziness or fainting

WHAT CAUSES IT
Inherited tendency present before birth

HOW IT IS DIAGNOSED
Echocardiogram, CT, or MRI

MAIN TREATMENT
Lifelong monitoring and valve or aortic procedures

CAN IT BE CURED?
No, valve anatomy cannot be reversed

IS IT AN EMERGENCY?
Sometimes, sudden severe chest or upper-back pain

LONG-TERM OUTLOOK
Varies widely with lifelong monitoring often needed

Overview

A bicuspid aortic valve is an aortic valve that formed with two flaps instead of the usual three. These flaps, also called cusps or leaflets, open and close to control blood flow from the heart's main pumping chamber into the aorta. The aorta is the large artery that carries blood to the rest of the body.

Bicuspid aortic valve is present from birth. It may work normally for many years, so some people do not learn they have it until adulthood. Others develop valve problems earlier in life.

Over time, a bicuspid valve may become narrowed or may not close tightly. Narrowing is called aortic stenosis . Leakage through the valve is called aortic regurgitation . Either problem can make the heart work harder.

Bicuspid aortic valve can also be associated with enlargement of the first part of the aorta, known as bicuspid aortic valve-related aortopathy. This matters because a significantly enlarged aorta may need closer monitoring or preventive surgery.

The effects vary widely from person to person. Some people need only periodic imaging. Others eventually need a procedure to repair or replace the valve or aorta.

Symptoms

Many people with a bicuspid aortic valve have no symptoms, especially when the valve works well and the aorta is not significantly enlarged. A clinician may first notice a heart murmur, which is an extra sound caused by turbulent blood flow.

When valve narrowing or leakage becomes more severe, symptoms can include:

Symptoms may develop gradually. A person might first notice that routine activities, such as climbing stairs or walking uphill, have become harder.

Contact your clinician promptly if you develop new exercise intolerance, breathlessness, chest discomfort, fainting, or swelling. These symptoms do not confirm a valve complication, but they should be evaluated rather than left until the next routine appointment.

Call emergency services for severe or sudden chest or upper-back pain, especially if it feels tearing or is accompanied by fainting, weakness on one side, trouble speaking, severe shortness of breath, or a cold sweat. These can be symptoms of an aortic emergency or another serious condition.

- Shortness of breath during activity or when lying down
- Chest pressure, tightness, or discomfort with exertion
- Unusual tiredness or reduced ability to exercise
- Dizziness, lightheadedness, or fainting
- A fast, pounding, or irregular heartbeat
- Swelling in the feet, ankles, or legs

Causes and risk factors

A bicuspid aortic valve develops before birth. It is not caused by exercise, diet, stress, or something a parent did during pregnancy.

The condition can run in families. A person may have a genetic tendency even when no specific

gene change is identified. First-degree relatives-parents, siblings, and children-have a higher chance of having a bicuspid valve or an enlarged aorta than people without this family connection.

Clinicians may recommend an echocardiogram for first-degree relatives, particularly when the affected family member also has enlargement of the aortic root or ascending aorta. An echocardiogram is a painless ultrasound examination of the heart.

Certain inherited conditions, including Turner syndrome, are associated with bicuspid aortic valve and aortic disease. However, most people with a bicuspid valve do not have Turner syndrome or another recognized genetic syndrome.

Factors that may affect the risk of complications include:

Normal aging can also lead to calcium buildup and stiffness in the valve. In a bicuspid valve, significant narrowing may occur at a younger age than it does in a typical three-leaflet valve.

- The amount of valve narrowing or leakage
- The size and growth rate of the aorta
- High blood pressure
- A family history of aortic dissection
- Coarctation, which is a narrowing in another part of the aorta
- Smoking and other cardiovascular risk factors

Diagnosis

A clinician may suspect a bicuspid aortic valve after hearing a murmur or an unusual clicking sound. Diagnosis requires imaging because a murmur alone cannot show the valve's shape or severity.

A transthoracic echocardiogram is usually the first test. A handheld device placed on the chest uses sound waves to create moving images of the heart. It can show:

Sometimes the valve or the entire aorta cannot be seen clearly on a standard echocardiogram. Computed tomography, or CT, can provide detailed pictures of the valve and aorta. Magnetic resonance imaging, or MRI, can also measure the aorta and assess blood flow without using ionizing radiation.

An electrocardiogram may be used to look at the heart's electrical activity. Exercise testing can help evaluate symptoms or exercise capacity in selected people, but it is not appropriate for every situation.

The clinician will interpret imaging measurements in context. Aortic size, body size, growth over time, family history, and other health conditions can all affect management. Comparing new images with earlier studies is particularly useful because it shows whether the aorta or valve disease is changing.

- Whether the valve has two leaflets
- How well the valve opens and closes
- The degree of stenosis or regurgitation
- The heart's size and pumping function
- Measurements of visible portions of the aorta

Treatment and management

A bicuspid valve that works normally may not require immediate treatment. Regular monitoring is still important because valve function and aortic size can change over time.

No medicine can change a bicuspid valve into a three-leaflet valve or reverse severe mechanical valve disease. Medicines may still be used to treat related conditions. For example, a clinician may prescribe treatment for high blood pressure to reduce stress on the heart and aorta. Cholesterol-lowering treatment is based on a person's overall cardiovascular risk, not simply on having a bicuspid valve.

Heart-healthy habits can support overall cardiovascular health. These include not smoking, maintaining a balanced diet, getting regular physical activity, and managing blood pressure, cholesterol, and diabetes when present.

Exercise advice depends on valve function, symptoms, blood pressure, and aortic size. Many people can remain active. People with significant aortic enlargement or severe valve disease may need to avoid intense straining or heavy lifting. A cardiologist can provide limits that fit the imaging findings and planned activity.

A procedure may be considered when stenosis or regurgitation becomes severe, causes symptoms, affects heart function, or meets other imaging criteria. Options can include:

Decisions about procedures are individualized. A heart valve team may include cardiologists, imaging specialists, and heart surgeons.

Bicuspid aortic valve alone is not usually a reason to take antibiotics like amoxicillin or doxycycline before dental work. Preventive antibiotics are reserved for people with specific high-risk heart conditions, such as certain replacement valves or a previous episode of infective endocarditis. Regular dental care remains important because bacteria from the

mouth can sometimes enter the bloodstream.

Pregnancy increases the amount of blood the heart must pump. People with significant valve disease or aortic enlargement should discuss pregnancy plans with a cardiologist and obstetric team before becoming pregnant when possible.

- Surgical valve repair: The surgeon reshapes the person's valve. Repair may be possible in selected cases, particularly for certain leaking valves.
- Surgical valve replacement: The damaged valve is replaced with a mechanical valve or a valve made from animal tissue. Each type has benefits and tradeoffs, including durability and the possible need for long-term blood-thinning medicine.
- Transcatheter valve replacement: A replacement valve is delivered through a blood vessel rather than through open-heart surgery. Bicuspid anatomy can make this more complex, so suitability depends on age, surgical risk, valve shape, calcium pattern, and aortic anatomy.
- Aortic surgery: An enlarged portion of the aorta may be replaced to reduce the risk of future tearing or rupture. This may be done alone or at the same time as valve surgery.

Complications

Potential complications include:

These complications are not inevitable. Scheduled imaging is intended to find important changes before they cause lasting heart damage or an aortic emergency.

- Aortic stenosis: A stiff, narrowed valve restricts blood leaving the heart.
- Aortic regurgitation: A valve that does not close fully allows blood to leak backward.
- Heart failure: Severe valve disease can weaken the heart or cause fluid buildup.
- Aortic aneurysm: Part of the aorta becomes enlarged.
- Aortic dissection: A tear develops within the layers of the aortic wall. This is uncommon but is a medical emergency.
- Infective endocarditis: Bacteria or other germs infect the valve or inner lining of the heart.
- Abnormal heart rhythms: Changes in heart size or pressure can contribute to rhythm problems.

Follow up

Follow-up usually continues throughout life, even if the valve works normally when it is first diagnosed. The timing of appointments and imaging depends on the amount of stenosis or leakage, aortic measurements, symptoms, and whether previous scans show change.

Keep copies of echocardiogram, CT, or MRI reports when possible. It can be helpful to know the size of the aorta, the severity of valve disease, and the date of the next recommended scan.

Tell your clinician between routine visits if you develop new breathlessness, chest discomfort, reduced exercise ability, fainting, palpitations, or leg swelling. Also discuss major changes such as pregnancy planning or starting a strenuous exercise or weight-training program.

Blood pressure should be checked regularly. Dental infections should be treated, and routine dental care should continue. Tell dentists and other healthcare professionals about any valve replacement, previous endocarditis, or aortic surgery because these details may change procedural planning.

First-degree relatives may wish to ask their own clinicians whether screening echocardiography is appropriate.

Sources

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