



For more information and to schedule an appointment with one of our cardiac specialists, please call:

615.222.HCM5 [4265]

PATIENT INFORMATION ON **Bicuspid Aortic Valve**

What is a Bicuspid Aortic Valve?

The aortic valve is a one way valve between the heart and the aorta. This valve is an exit only door to the aorta. Normally, the aortic valve has three small flaps or leaflets that open widely and close securely to allow the blood to flow from the heart to the aorta—preventing blood from flowing backward into the heart.

In the case of a Bicuspid Aortic Valve (or BAV), the valve is deformed and there are only two functional leaflets. This deformity occurs at birth. Although, the valve may not function perfectly, it may function adequately for years without causing any symptoms or signs of a problem.

Why is this important?

With time and research, it has been suggested that bicuspid aortic valve disease is caused by a disorder of the connective tissues. It can also cause circulatory system problems like problems with the coronary arteries, aortic aneurysms or the thoracic aorta.

What causes a Bicuspid Aortic Valve?

The actual causes of a bicuspid valve remain unclear. It is known however, that a two leaflet valve develops in the early stages of pregnancy and the defect is present at birth. This is a genetic condition that can be passed on to first degree relatives and does require testing of families as a part of complete treatment. Genetic counseling prior to pregnancy is also strongly encouraged.

What are the symptoms of Bicuspid Aortic Valve disease?

If a bicuspid aortic valve is present, it has been there since birth. Most people have this condition diagnosed as an adult because the bicuspid valve can function for years without any problems. Typically, patients will have had a long standing childhood murmur. As the patient ages, the valve may be more prone to symptoms or problems.

Valve problems that arise from a bicuspid aortic valve are typically aortic stenosis or aortic regurgitation. As the valve ages, it is more likely for calcium to form around the leaflets. When this occurs, it can make the aortic valve opening stiff and narrow. This causes a condition called aortic stenosis. This makes it more difficult for the blood to be pushed through an already dysfunctional aortic valve. The heart has to pump harder to force blood through the valve. Symptoms of aortic stenosis include:

- **Chest pain or Angina**
- **Dizziness**
- **Fainting or Syncope**
- **Shortness of breath** [Especially with Activity]

When the bicuspid aortic valve does not close completely, it is called aortic regurgitation. When this occurs, the blood can flow backwards to the heart. The heart must then pump harder in order to pump that same blood out. Over time, this can cause strain on the heart's pumping chamber (the left ventricle), and become dilated, thickened and over expanded. Symptoms of aortic regurgitation are:

- **Shortness of breath** [Especially with Activity]
- **A sensation of the heart “pounding” against the chest wall that causes an awareness of heartbeat**
- **Chest pain or Angina**
- **Palpitations** [feeling like the heart is skipping a beat]

Due to the extra force required to pump blood through the diseased valve, heart failure can occur. Once the ventricle is enlarged, it cannot pump blood efficiently and can lead to signs and symptoms of heart failure.

There are studies to suggest that a bicuspid aortic valve is caused by a connective tissue disorder. This connective tissue disorder also affects the lining of the blood vessels in the body including the aorta. This can increase the chance of developing an aneurysm. As this occurs, the walls of the vessel begin to thin and deteriorate. The walls may become weak, thin and unstable which could in turn lead to an aneurysm. The inner or outer layers of the aneurysm can rupture causing a dissection. This can be a life threatening emergency. Aneurysms can also occur and be stable for many years and never rupture. Any type of aneurysm discovered will require regular follow up and monitoring to assure its stability. Even with monitoring, a large or troublesome aneurysm will require surgical intervention if a threat is determined.

There is a 40% incidence of a thoracic aneurysm in patients with a bicuspid aortic valve. Also there is a 30% incidence of this disorder in 1st degree relatives. Family members will either inherit the BAV itself or thoracic aneurysms. Because of this, routine follow ups and screenings are strongly encouraged among patients and families of those diagnosed with bicuspid aortic valve disease.

As mentioned earlier, many people that have bicuspid aortic valve disease do fine with little to no signs or symptoms of the condition. It should be mentioned that about 30 - 50% of patients with BAVD have complications. These complications can range from very minor to life threatening. This is the driving force behind the push for ongoing care and medical management by a specialist in heart valve diseases. A trained cardiac specialist can monitor changes in the aorta, heart and valves regularly and over an extended period of time.

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What is the treatment for a Bicuspid Aortic Valve?

Some people can go their entire lives and have a bicuspid aortic valve and never know it. In other cases, it may have to be treated with medications and/or surgical interventions to prevent other conditions that make having BAVD more dangerous. Hypertension is an important condition that must be treated aggressively in the presence of BAVD. If a patient is a smoker, it is critically important that they stop immediately.



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