Children's Heart Center of Central Oregon

# Bicuspid aortic valve

****The following is general information about bicuspid aortic valve (BAV). However, please keep in mind that every case is unique, and requires different follow-up, testing, treatment, and restrictions. Please speak with your cardiologist about the particular recommendations for your child.****

## *What is a bicuspid aortic valve (BAV)?*

The aortic valve is the valve that blood passes through to get from the heart to the aorta (the main vessel that supplies the body with blood). The aortic valve normally has three leaflets. If the valve is bicuspid, it means that there are only two leaflets. See http://www.pted.org/?id=bicuspidvalve1 for a diagram.

## *How common is BAV?*

BAV is the most common congenital heart defect. It is present in 1-2% of the population.

## *What are the potential complications of having a BAV?*

When a valve is bicuspid, it may not function normally. Bicuspid valves can develop aortic stenosis, which means that the valve may not open as well as it should, causing some obstruction to blood flow. Bicuspid valves can also develop aortic insufficiency, which mean that the valve does not close as well as it should, causing reverse flow of blood, or a "leak," back into the heart.

Patients with bicuspid aortic valve can also develop dilation (enlargement) of the aorta. If the enlargement becomes severe, there is a risk of tearing of the aortic wall. Therefore, it is important to know whether there is any enlargement present.

Finally, children with BAV can have other heart defects as well. The most common associated heart defect is coarctation of the aorta, which is a narrowing of the aorta.

## *Can BAV be fixed?*

If any of the above complications becomes severe or is causing symptoms for the child, repair can be performed using a variety of surgical or heart catheterization procedures. The specific procedure depends on each patient's unique circumstances. For example, children with severe aortic stenosis can be treated with a balloon valvuloplasty procedure, which is performed using a catheter and doesn't require opening the chest. On the other hand, children with severe aortic insufficiency and/or severe aortic dilation will likely require an open heart operation to repair or replace the valve or the part of the aorta that is dilated.

***How is BAV diagnosed?***

In most cases, BAV can be diagnosed with an echocardiogram, which may be ordered because a murmur is heard or because there is a family history of BAV. Aortic stenosis, aortic insufficiency, and aortic dilation can also be seen on an echocardiogram.

***What kind of follow-up should I expect if my child has a bicuspid aortic valve?***

Children with BAV need lifelong follow-up with a cardiologist. Complications from a BAV are highly variable, where a BAV can function normally (i.e. no stenosis, insufficiency, aortic dilation, or symptoms), or may have severe abnormalities requiring intervention soon after birth. Even if a BAV has normal function when the child is young, the valve can change over time. Therefore, regular echocardiograms to monitor the aortic valve and the aorta are very important, and help determine when and if it is time for intervention with medication, surgery, or sports restriction.

***My child has a BAV. Can he/she play sports?***

Many children with BAV have no sports restrictions. However, some children, particularly those with significant stenosis, insufficiency, or aortic dilation, should be restricted from competitive sports, and/or activities with a high isometric component. These are activities that involve tightening of muscle groups, which can cause very high blood pressure and can have a detrimental effect on the aortic valve and the aorta. There are many types of isometric sports, but common examples include weight lifting, wrestling, football, and gymnastics. Please speak with your child's cardiologist to learn if your child should be restricted from any activities.

***Is there treatment for aortic dilation?***

Some degree of aortic dilation ("aortopathy") happens in 50-90% of patients with BAV. This can progress over time, and is one reason why echo monitoring is so important. Aortic dilation does not typically cause a heart murmur or symptoms, so imaging (echo, CT, or MRI) is the only way to know that it is present. If aortic dilation becomes progressive, medications can be used to try to slow its progression. These medications include beta blockers (e.g. atenolol) or angiotensin receptor blockers (ARB's, such as losartan or irbesartan). Speak with your cardiologist about whether your child would be a candidate for any of these options. In addition to BAV, there are multiple other genetic causes of aortopathy (see https://marfan.org for more information). If your child has significant aortic dilation, your cardiologist may recommend genetic testing to rule out other genetic causes.

***What are the genetics of BAV?***

The genetics of BAV are not fully understood, but we know that there is a very strong genetic component. Studies have shown that BAV is present in about 9% of first degree relatives (parents and siblings) of patients with BAV. For this reason, it is recommended that *all first degree relatives of a person with BAV be screened for the condition*. Screening is typically performed using an echocardiogram.