BICUSPID AORTIC VALVE DISEASE

Northwestern’s Bluhm Cardiovascular Institute
Center for Heart Valve Disease

Martha and Richard Melman Family Bicuspid Aortic Valve Program
What is Bicuspid Aortic Valve?

The aortic valve controls the flow of blood from the heart to the aorta, and out to the body.

Normally, the aortic valve has three leaflets that work to keep blood moving in one direction. With bicuspid aortic valve (BAV), two of the three leaflets of the aortic valve fuse during a person’s fetal development. This creates a bicuspid or two leaflet valve, instead of the normal tricuspid or three leaflet valve. BAV also affects the thoracic aorta, the largest artery in the body that transports blood from the heart to the body.

Present at birth, BAV is one of the most common congenital heart disorders, affecting about one to two percent of the population. BAV often runs in families. It is at least twice as likely to occur in men as in women.

A heart with BAV may work normally without causing any symptoms for the person. However, many people will develop complications from their BAV and may need surgery in their lifetime - either to fix the aortic valve, the thoracic aorta or both.

While a heart with BAV may work normally without causing any symptoms, BAV puts strain on:

- The aortic valve
- The heart
- The thoracic aorta

Symptoms and Diagnosis

Symptoms of BAV are caused by an aortic valve that leaks or does not open completely. Symptoms of BAV may include:

- Chest pain
- Shortness of breath
- Racing heartbeat
- Fainting

One of the earliest indicators of BAV may be a heart murmur. A heart murmur is an abnormal sound caused by the turbulent flow of blood over a diseased heart valve.

A cardiologist will order an echocardiogram (ECHO) to determine whether you have BAV. An ECHO is a painless test that uses sound waves to create a moving picture of the heart. It is the most accurate way of diagnosing BAV.
If you are diagnosed with BAV, you should receive ongoing care from a cardiologist with experience in heart valve disease.

Medical Care and Testing

If you are diagnosed with BAV, you should receive ongoing care from a cardiologist with experience in heart valve disease. A cardiologist will carefully monitor any changes in your heart function, valves and aorta over time.

Cardiologists with expertise in heart valve disease will use advanced imaging tests to assess your heart's condition over time. Because the time between diagnosis of BAV and the need for potential surgery is different for everyone, it is important to be seen by a cardiologist, as recommended, for follow-up care. Follow-up care allows your cardiologist to monitor your BAV, aorta and overall heart function by using a variety of advanced imaging tests.

ADVANCED IMAGING TESTS USED TO MONITOR BAV

- Echocardiogram (ECHO): a test that uses sound waves to create a moving picture of the heart
- Computed Tomography Scan (CT Scan): a test that combines X-ray and computer technology to produce cross-sectional images of the bones and soft tissues in your body
- Cardiac Magnetic Resonance Imaging Scan (MRI): a test that uses pulsed radio frequency and magnets to create images of the heart
- Magnetic Resonance Angiography (MRA): a test that uses pulsed radio frequency and magnets to detect problems with blood vessels and blood flow
- 4-D MRI Scan: cutting edge technology used and developed at Northwestern and other select hospitals offering the most advanced non-invasive views of your heart and aorta

Family Screening

If you are diagnosed with BAV, your family members have a nine percent chance of having BAV as well. This makes family screening very important because the majority of people with BAV have no symptoms until they begin to have complications.

If you have been diagnosed with BAV, the American College of Cardiology/American Heart Association recommends BAV screening for your first-degree family members, which include:

- Parents
- Siblings
- Children

PREGNANCY

If you are a woman with BAV and want more information about pregnancy and BAV, you should contact Northwestern’s Heart Disease and Pregnancy Program. This program provides specialized care for pregnant women with heart disease. Marla A. Mendelson, MD, the program’s founder and director, is known for her research on:

- Heart disease during pregnancy
- Heart disease in women
- Adult congenital heart disease

It is important for all women of childbearing age who have heart disease and are considering pregnancy to receive a complete preconception health evaluation prior to becoming pregnant.

Please call 312-NM-HEART or visit us at heart.nmh.org to speak with us about our Heart Disease and Pregnancy Program or to schedule an appointment.
Complications from BAV

Complications associated with BAV can be serious and life-threatening. This is why diagnosis and ongoing care by a cardiologist is important.

Complications of BAV may include:

- Aortic regurgitation/insufficiency: the aortic valve does not close completely, causing the valve to leak
- Aortic stenosis: the aortic valve does not open wide enough, causing restricted blood flow
- Infective endocarditis: infection of the lining of the heart chambers and the heart valves
- Aortic aneurysm: ballooning of the wall of the aorta
- Aortic Dissection: bleeding into and along the wall of the aorta
- Coarctation of the aorta: narrowing of the aorta at some point along its length

Treatment for BAV

Once you are diagnosed, your treatment options depend on whether or not you have symptoms.

If you have no symptoms

Medical management is used to treat most people with BAV who do not have symptoms. Medical management can help:

- Slow the onset of complications
- Prevent further complications
- Recognize the onset of symptoms
- Identify optimal timing of surgical intervention

Medical Management

The goal of medical management of BAV is to achieve optimal blood pressure, heart rate and cholesterol levels. You and your cardiologist will decide if and when medications are necessary.

You can also help manage your BAV by being heart healthy. Ways to be heart healthy include following a low fat, low sodium diet, being physically active, practicing good dental hygiene and quitting or avoiding smoking.
Follow-up Routine Testing

Even for those people with BAV who have no symptoms routine, repeated imaging tests are important. These tests will assess any changes in your BAV and heart function. These tests may also detect possible complications that can arise in the aortic valve and thoracic aorta. Your cardiologist will tell you when to have follow-up testing and which test(s) you will need.

If you have symptoms

While medical management of your BAV will continue, once your aortic valve starts to leak or does not open completely, surgery may be your best treatment option.

Surgery

Sometimes cardiac surgeons can fix BAV by performing an aortic valve repair. However, if the aortic valve is too damaged to repair, an aortic valve replacement may be the best option. You may also require surgery to repair or replace sections of your aorta. When it is determined that surgery is needed your cardiac surgeon will discuss which option(s) is best for your particular needs.

Postoperative Long-Term Care

Even after you have surgery on your BAV, you may be at risk for future problems with your aorta. Because of this, it is important to continue follow-up care with your cardiologist, as they will create a plan of care to ensure that you are monitored appropriately.

How We Help People with BAV

Enroll in the Martha and Richard Melman Family Bicuspid Aortic Valve Program

Our unique program offers comprehensive care for people with BAV and their first-degree relatives. Our cardiologists and cardiac surgeons offer highly sophisticated treatment options for people with BAV ranging from medical therapy to highly advanced surgical interventions.

The Martha and Richard Melman Family Bicuspid Aortic Valve Program offers:

• Expert clinical care from board-certified cardiologists and cardiac surgeons, specializing in heart valve disease
• Coordination of care with your local physicians including providing updates on your care and progress
• Monitoring and evaluation of your BAV through the use of ECHO, CT scan, and/or MRI testing
• Advanced diagnostic 4-D MRI Scan giving highly detailed and advanced non-invasive images of your valve and aorta
• An invitation for your first-degree relatives to participate in the program due to the hereditary nature of BAV
• Access to ongoing clinical research specifically for BAV

The Martha and Richard Melman Family Bicuspid Aortic Valve Program is co-directed by:

• Jyothy Puthumana, MD, a cardiologist specializing in medical treatment for BAV
• S. Chris Malaisrie, MD, a cardiac surgeon specializing in surgery for BAV
Drs. Puthumana and Malaisrie collaborate with Patrick M. McCarthy, MD, an internationally known cardiac surgeon who specializes in valve surgery, and Robert O. Bonow, MD, a cardiologist who is an internationally renowned expert on valve disease and the past chair of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee on Management of Patients With Valvular Heart Disease).

Together, these experts lead a team of:

- Cardiologists
- Cardiac surgeons
- Cardiac Radiologists
- Nurses, including a dedicated BAV nurse coordinator
- Researchers
- Cardiac rehabilitation specialists
- Psychologists
- Social workers

Colleen A. Clennon, RN, BSN, is the BAV nurse coordinator for the Martha and Richard Melman Family Bicuspid Aortic Valve Program. With the goal of personalized treatment to meet individual needs of people with BAV, Colleen will help to coordinate testing and treatment and answer any questions that you may have.

Please contact Colleen at bav_rn@nmh.org or 312-NM-HEART (664-3278).

**Second Opinions**

Our cardiologists and cardiac surgeons are available for second opinion consultations on your medical and surgical treatment options for BAV. Please contact Colleen at bav_rn@nmh.org or 312-NM-HEART (664-3278). For more information, visit nm.org/secondopinion.
Northwestern Medicine and Bluhm Cardiovascular Institute

Recognized for providing exemplary patient care and state-of-the-art advancements.

Northwestern Medicine® is the collaboration between Northwestern Memorial HealthCare and Northwestern University Feinberg School of Medicine around a strategic vision to transform the future of healthcare. It encompasses the research, teaching, and patient care activities of the academic medical center.

Sharing a commitment to superior quality, academic excellence and patient safety, the organizations within Northwestern Medicine comprise a combined workforce of more than 25,000 among clinical and administrative staff, medical and science faculty and medical students.

The entities involved in Northwestern Medicine remain separate organizations. Northwestern Medicine is a trademark of Northwestern Memorial HealthCare and is also used by Northwestern University.

Northwestern Medicine ranks #11 in the U.S. News & World Report 2015-16* Honor of America’s Best Hospitals. The hospital is recognized by U.S. News & World Report in 13 of 16 clinical specialties in #1 Illinois and Chicago in their state and metro rankings, respectively.

Northwestern Medicine has nursing Magnet Status, the nation’s highest recognition for patient care and nursing excellence. For 15 years running, Northwestern Medicine has been rated among the “100 Best Companies for Working Mothers” guide by Working Mother magazine. The hospital is a recipient of the prestigious National Quality Health Care Award and has been chosen by Chicagoans as the Consumer Choice according to the National Research Corporation’s annual survey for 15 years.

The Bluhm Cardiovascular Institute is a world-class heart and vascular program that is a destination for those requiring highly specialized cardiovascular care. Our clinical excellence, comprehensive programs and superior clinical outcomes have again been recognized at the national level.

In 2015-16 U.S. News & World Report ranked our Cardiology and Heart Surgery program #9 in the nation for the last eight years, the top ranked program in Illinois and the region.

*U.S. News & World Report, America’s Best Hospital, 2015-16