

Northwestern launches comprehensive program for patients with bicuspid aortic valve disease

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Bicuspid aortic valve (BAV) disease is the most common congenital heart defect, occurring in approximately one to two percent of the population. The condition is present when the aortic valve, the valve that connects the heart to the main artery that distributes blood to the body, contains two leaflets instead of three, which open and close to regulate blood flow. As a result, the valve does not function properly, which can cause strain on the heart over time and may lead to serious health complications. Northwestern Memorial Hospital's Bluhm Cardiovascular Institute (BCVI) has been at the forefront of BAV research and treatment for years and recently launched a comprehensive program that brings together a multidisciplinary team of specialists experienced in the diagnosis and medical and surgical management of patients with BAV.

"Care for [patients](#) with bicuspid aortic valve is complex," said Patrick McCarthy, MD, director of Northwestern's Bluhm Cardiovascular Institute. "This program is the first to support a patient from A to Z, through screening, monitoring, family assessment, treatment and research."

One of the complications of BAV is when [calcium deposits](#) prematurely form on the valve flaps, causing the valve to stiffen and the opening narrows. This condition is known as aortic stenosis and may lead to shortness of breath, chest pain and blackout spells. A leaky valve may not close securely, causing blood to flow backwards into the heart. This

[regurgitation](#) causes the heart to overwork, and the [ventricle](#) may enlarge causing [heart failure](#). BAV can also be associated with ballooning of the aorta as it arises from the heart. Aneurysms of the ascending aorta can lead to life-threatening rupture if not treated in a timely fashion.

"Close monitoring of BAV patients is imperative in order to prevent life-threatening complications," said Jyothy Puthumana, MD, cardiologist and medical director of the Bicuspid Aortic Valve Program at the Bluhm Cardiovascular Institute. "By bringing together a team of specialists, we are able to provide patients with comprehensive care beginning at diagnosis and can carefully anticipate when an intervention may be needed."

Cardiologists, cardiac surgeons, radiologists and a dedicated research nurse work together to coordinate care for patients. "My role is to make sure each patient has a seamless journey with us," said Colleen Clennon, RN, Bicuspid Aortic Valve Nurse Clinician at the Bluhm Cardiovascular Institute.

"By using state-of-the-art research techniques, our research team is focused on providing new knowledge that will help guide patient care and improve outcomes for patients and their families with BAV," said Paul Fedak, MD, PhD, Adjunct Associate Professor at the Northwestern Feinberg School of Medicine. "Our rapidly expanding tissue and blood bank will allow our team to explore the molecular and cellular mechanisms that cause the changes that complicate BAV. Our integrated scientific and clinical approach will apply this knowledge and allow our team of physicians to better manage this common and sometimes challenging problem."

The cause of BAV is not completely clear, but experts believe it develops during the early stages of pregnancy. It can run in families with an estimated ten percent of cases attributed to hereditary causes.

"Our program extends care to first degree relatives who may also be impacted by BAV," added Puthumana. "Through screening family members of patients with BAV, we can identify cases of BAV earlier and begin monitoring patients before problems present."

Symptoms associated with BAV can include chest pain, shortness of breath, a racing heartbeat and fainting spells due to a restricted valve. While some patients will live for years without symptoms or any sign of concern, an estimated 60 percent will require surgical intervention at some point in their lifetime.

When BAV is suspected either due to the presence of a heart murmur, which is often the first sign, or because of family history, doctors rely on an echocardiogram – ultrasound of the heart to look at the aortic valve and the [ascending aorta](#). BCVI doctors also have access to 4D flow MRI, a novel imaging technique that offers a more comprehensive picture of the [heart](#) and the aorta. The advanced imaging led by Michael Markl, PhD, director of cardiovascular MRI research at BCVI, and his team, allows clinicians to better visualize blood flow through the cardiovascular system and potentially identify higher risk imaging markers by MRI that would warrant closer follow up.

"The decision to proceed with surgery is an important one and should be determined by a team of specialists," said Chris Malaisrie, MD, surgical director of the Bicuspid [Aortic Valve](#) Program at BCVI. "With timely surgery, life-threatening complications from [bicuspid aortic valve](#) can be prevented."

Provided by Northwestern Memorial Hospital

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