





Bicuspid aortic valve

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Bicuspid aortic valve (BAV) is the most common congenital heart disease with an estimated prevalence between 0.5% and 2%¹. BAV is usually made of 2 unequal-sized leaflets where the larger leaflet has a central raphe or ridge that results from fusion of the commissures. Most common is the fusion of the right and left cusps which is associated with coarctation of the aorta. Pure BAV is rare, and it has symmetrical leaflets or there is no raphe². The majority of BAV are an isolated birth defect, but there are also genetic causes. It can appear as a part of a syndrome (e.g., Shone's syndrome, Turner syndrome, Williams syndrome, Anderson syndrome), or because of a single gene mutation such as *NOTCH1* and *ACTA2*, or a combination of multiple gene mutations. Recently, the role of the extracellular matrix role in the differentiation of the cells and formation of the leaflets is being investigated^{1,2}. Because 20-84% of patients with a BAV develop ascending aortic dilatation, BAV should be regarded as a valvulo-aortopathy. The dissection incidence in BAV patients is eight times higher than in the general population³. The clinical presentation of patients with BAV varies from mild to severe valve disease and the symptoms typically develop in adulthood. The clinical manifestations depend on the function of the aortic valve, the aortopathy and acquired complications such as endocarditis. Infectious endocarditis affects 10-30% of BAV patients during their lifetime and is more common in younger male patients¹. Due to abnormal hemodynamics on the aortic wall and different activity of the matrix metalloproteinases, BAV is associated with dilatation in the aorta, especially the aortic root and ascending aorta, and coarctation of the aorta often occurs simultaneously. With the incidence between 59% and 81%, aortic valve stenosis is the most common complication of the BAV, with calcification playing a major role^{1,2}. Treatment strategies vary depending on the severity of the disease and dilation of the aorta. Together with valve replacement, transcatheter aortic valve implantation is now considered as an alternative option with good results¹⁻³.

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LITERATURE

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