Prevalence of Congenital Heart Diseases has increased in the last decades in line with certain treatment and surgical interventions available. Bicuspid aortic valve with coarctation of aorta is aortic vasculopathy with three main approaches you need to be familiar with:

Both BAV and CoA can be present together and most commonly associated with aortic or mitral valve stenosis and malformation (Ref-JHD78F). The main focus of this review is on etiology, epidemiology, pathophysiology, evaluation and diagnosis, treatment as well as postoperative complications and prognosis.

Bicuspid Aortic Valve (BAV) is an aortic valve with two cusps making it difficult to pass and pump blood leading to aortic stenosis. It is most commonly associated with an underlying narrowing of a part of the aorta called Coarctation presented at birth.

### Etiology and epidemiology

During fetal development there is abnormal migration of neural crest cells and absence of NO Synthase which causes abnormal fusion of cusps of aorta resulting bicuspid valves. This leads to abnormal blood flow due to aortic stenosis and coarctation of aorta most commonly at a region of Patent Ductus Arteriosus.

Coarctation of aorta is prevalent among ~7% of patients with CHD and ~0.07% in the general population. Bicuspid aortic valve disease is found 1-2% in the general population. CHD can also be found in association with certain genetic diseases like Turner and Williams-Beuren Syndrome (Ref-A1B2C3).

### Pathophysiology

The left and right coronary cusp fusion is the most known cause of Bicuspid Aortic Valve, this abnormality is linked to aortic coarctation. In the presence of redundant tissue, valvular incompetence is the most common disease. The newborn with coarctation while PDA is closing can preserve perfusion to lower extremities.

PDA is less likely to contribute unless the remaining left heart is hypoplastic in Lower-extremity Pulse Oximeter Screening in newborns. Therefore, a baby might often pass with an adequate saturation. In older children aortic-coarctation causes upper extremity hypertension, which can progress to Aortic Aneurysm, Coronary Artery Disease (CAD), Cerebrovascular anomaly.

The diagnostic approach of BAV with CoA is linked to history, including signs, symptoms and physical examination as well as various routine evaluation techniques.

### Post-operative complications

Life-long follow up is required even after surgery to avoid life-long risk of following post-operative complications:

### Prognosis

Aortic sinuses or ascending aorta surgery is required in 30% of bicuspid aortic valve patients in addition to aortic valve replacement (Ref-DJ49F2). According to current AHA guidelines, if a surgeon replaces an aortic valve for whatever reason and the size of the aortic root is larger than 4.5 cm, the aortic root should also be replaced.

### Differential diagnosis

The idea of the article is to highlight various treatment options available for above Congenital Heart Disease and its aspects in terms of valvulopathy, aortopathy and its’ link to heredity.