GEN19 - Predictors of Aortic Dissection in Patients with Bicuspid Aortic Valve Disease

OBJECTIVE: To assess data from the GenTAC registry in an effort to determine if clinical, imaging or genetic variables can help clinicians assess the risk of an individual patient with BAV dissecting.

ORGANIZATION

Lead Investigator: Anna M Booher, MD
Co-Investigators: Kim Eagle, MD
Funding Source: GenTAC

BACKGROUND AND RATIONALE

Bicuspid aortic valve (BAV) disease affects an estimated 1-2% of the population and is the most common congenital abnormality that presents in adulthood. The aortic tissue in patients with BAV has been shown to have reduced elastic properties compared with normal aortas. The proposed cellular mechanism for this abnormality is that there is a developmental defect of neural crest cells resulting in early vascular smooth muscle apoptosis, leading to decreased production of tissue inhibitors of matrix metalloproteinase and subsequent increased degradation of fibrin and elastin.

The sequelae of aortic dilation, although rare, can include the catastrophic consequences of dissection or rupture; and can unfortunately be the first clinical manifestation of the disease. Although an area of ongoing study, the clinical variables that predict aortic catastrophes in patients with BAV are not well elucidated.

DESIGN

Hypothesis: Patients with BAV who dissect may have a different clinical profile from those who do not dissect

Inclusion criteria: Subjects who have a bicuspid aortic valve.

Samples: None

Data: Clinical evaluation, imaging, genetic and surgical data.

RESULTS: Results pending