

PATTERNS OF AORTOPATHY AND VALVE PATHOLOGY DIFFER IN CHILDREN AND YOUNG ADULTS WITH BICUSPID AORTIC VALVE AND MARFAN SYNDROME

BACKGROUND: Patients with bicuspid aortic valve (BAV) and patients with Marfan syndrome (MFS) are both at increased risk for aortic dilation and dissection. The purpose of this study is to compare patterns of aortopathy, aortic valve pathologies and rates of aortic surgical intervention in patients with either BAV or MFS.

METHODS: We performed a retrospective review of all patients older than 10 years of age who presented to our center with either BAV or MFS from 1990 to 2014. Moderate or greater aortic stenosis (AS) was defined as a valve gradient >3.5 m/s; aortic insufficiency (AI) was quantified using standard criteria. Aortic diameter was measured at 4 levels, and Z-scores were computed. Freedom from aortic root repair or replacement was determined using long-rank calculations.

RESULTS: Four hundred thirty-two patients were identified: BAV n=358, MFS n=74. Median age was 20 years (10-40), with patients with BAV being younger (BAV, 17.5 yrs; MFS, 21.5 yrs; $p<0.001$). Moderate or greater AS and moderate or greater AI were observed most often in patients with BAV. Conversely, dilation of the aortic root or ascending aorta was more common in patients with MFS. The two patient populations had different patterns of aortic dilatation. Patients with MFS had greater dilation, as measured by Z-scores, of the sinus of Valsalva and sinotubular junction while patients with BAV had greater enlargement of the ascending aorta. Valve-sparing aortic root replacement (David, Yacoub) was performed in 12 patients (BAV, n=7; MFS, n=5) and aortic root replacement (Bentall, Ross) was performed in 45 patients (BAV, n=34; MFS, n=11). Freedom from surgical intervention on the aortic root at 30 years of age was significantly better in the BAV group (BAV, 92% vs. MFS, 80%; $p=0.003$).

CONCLUSIONS: Patients with BAV and MFS have different patterns of aortopathy and AV pathology. Dilation of the ascending aorta and AV pathology is more common in BAV while dilation of the aortic root is seen more frequently in MFS. Our data may help inform disease specific patient counseling and follow up.