

Natural History of Bicuspid Aortic Valve Disease Over First Two Decades of Life

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Introduction: The natural history of bicuspid aortic valve (BAV) has not been elucidated in pediatric population. Our aim was to study the age wise changes in various echocardiographic parameters in patients with BAV over first two decades of life.

Methods: The Mayo Clinic echocardiography database was retrospectively analyzed to identify pediatric and young adult subjects (≤ 22 years) diagnosed with BAV from 1990-2015. We analyzed all echocardiograms performed prior to any intervention on aortic valve or aorta and recorded measurements formatted as z-scores.

Results: We identified a total of 1010 patients with BAV, out of which 619 had isolated BAV. The distribution of morphology of cusp fusion in isolated BAV was right-left (RL) in 419 (67.7%), right-non coronary fusion (RN) in 197 (31.8%) and left-non coronary fusion (LN) in 3 (0.5%) subjects. There was an abnormal dilation of mid-ascending aorta (defined as z-score > 2) during pediatric and young adult age. Subjects with RN fusion had significantly higher dilation than patients with RL fusion ($p < 0.001$). There was a period of peak growth of aorta around 8 to 10 years of age present only in the males. There was no significant sinus of Valsalva dilation. There was a higher mean aortic valve systolic gradient at birth due to congenital aortic stenosis but no significant progression was seen over time. However, aortic regurgitation showed age wise progression with higher grades in patients with RN cusp fusion ($p < 0.001$).

Conclusions: The natural history of BAV shows interesting patterns of disease progression in terms of growth of aorta and aortic valve abnormality over time based on valve morphology.

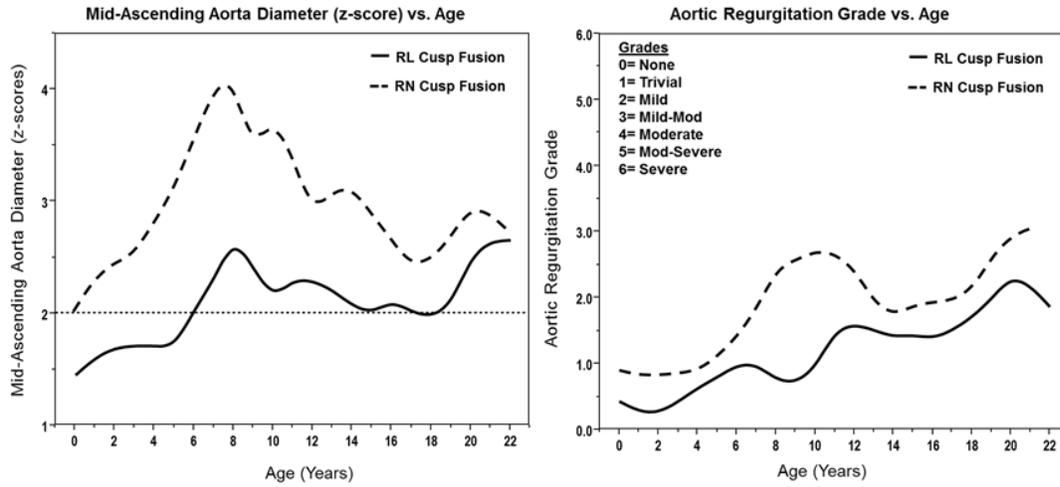


Figure.1 Age wise progression of aortic diameter and aortic regurgitation grades in patients with BAV. Age in years is plotted along x axis. Mid-Ascending aortic dimensions as z-scores and aortic regurgitation as grades are plotted along y axis. Solid line represents RL and dashed line represents RN cusp fusion. P-values represent the difference between means of RL and RN cusp morphology in each category.