Very low progression rate of ascending aorta dilatation in children with isolated bicuspid aortic valve

Division of pediatric and fetal cardiology, Department of pediatrics, Centre de Recherche du Centre Hospitalier de Universite de Sherbrooke and University of Sherbrooke, Sherbrooke, Canada (1), Department of general surgery, Maisonneuve-Rosemont Hospital, Montreal, Canada (2), Division of pediatric cardiology, Department of pediatrics, Centre Hospitalier Universitaire Sainte-Justine, Montreal, Canada (3)

Introduction: Bicuspid aortic valve (BAV) is the most common congenital heart disease. Children with BAV have an increased risk of developing aortic valvar disease and progressive ascending aorta dilatation. Studies have linked aortic dilatation to the severity of valvar disease and authors have hypothesized that dilatation is caused by increased shear stress on the aortic wall. Some studies have also reported distinct dilatation patterns according to the type of leaflet fusion. However, we are still unable to predict which children with BAV will present a significantly dilated ascending aorta when entering adulthood.

Objectives: We sough to determine longitudinal risk factors of progressive ascending aorta dilatation in children with BAV and to evaluate the risk of dilatation in patients with isolated BAV (without significant stenosis or regurgitation).

Methods: We extracted all echocardiography reports performed on BAV patients aged <20 years old between 1999 and 2016 at our institution. We collected information on type of fusion, severity of valvar disease, presence of coarctation of the aorta and ascending aorta diameter (normalized to Z-scores). We used multivariate linear mixed models to evaluate the influence of hemodynamic risk factors and BAV type on aortic dilatation rate and then estimated the probability of severe dilatation at 18 years of age according to different hemodynamic profiles.

Results: A total 761 patients (3,148 echocardiograms) were included. Median follow-up was 4.4 [IQR 1.0-8.4] years. Increased aortic dilatation rate was associated with severity of aortic regurgitation and severity of aortic stenosis. In patients with isolated BAV, aortic dilatation rate was low (0.05 Z-score unit per year) and showed no association with fusion type. The estimated probability of developing a dilated ascending aorta with a Z-score > 8 at 18 years of age was 1.0% in patients with isolated BAV and 9.5% in patients with at least moderate stenosis and regurgitation (figure 1). We observed no dissections or surgery for reduction of aortic size during follow-up period.

Conclusions: In children with isolated BAV, the mean rate of aortic dilatation is low but significantly increases in presence of valvar stenosis and regurgitation and is not associated with BAV type.

Figure1: Predicted ascending aorta Z score at 18 years old