Increased aortic stiffness in prepubertal girls with Turner syndrome.


(1) Ghent University Hospital, Ghent, Belgium
(2) Ghent University, Ghent, Belgium

Introduction: aortic dilation and dissection contribute highly to the increased mortality of Turner syndrome (TS) but the exact pathophysiology is not completely understood.

Methods: we investigated aortic diameters and aortic wall properties in young prepubertal TS girls. Fifteen prepubertal TS girls (median age 10.64, IQ 8.31-11.04) with a tricuspid (TAV n=9) or a bicuspid (BAV n=6) aortic valve, and 31 sex, age and height matched healthy controls underwent a cardiac and vascular ultrasound to evaluate aortic dimensions and elastic properties of the aortic wall.

Results: TS BAV had significantly larger ascending aortic diameters than controls for absolute diameter 22.2±5.10mm vs 18.7±1.92mm (p=0.017) and z-score 1.8±2.05 vs 0.2±0.73 (p=0.004). Distensibility of the ascending aorta was lower in the TS than in controls (40.2 10-3kPa-1, IQ 31.3-56.2 vs 62.9 10-3kPa-1, IQ 55.5-76.5, p=0.002) both for TS TAV (p=0.012) and BAV (p=0.003). Stiffness index was higher in TS than in controls (5.26 , IQ 3.34-5.26 vs 2.55-3.24, p=0.004) both for TS TAV (p=0.027) and TS BAV (p=0.004). Pulse wave velocity along the whole aortic arch was not different between groups. There was no correlation between stiffness and z-score of the ascending aortic diameter.

Conclusions: we concluded that in prepubertal TS girls stiffness of the ascending aorta is increased in patients with a BAV and TAV while dilation of the ascending aorta is more frequent in BAV. This suggest an intrinsic aortic wall abnormality making all TS patients at increased risk for severe aortic complications although the risk is the highest for TS with BAV.