

**Evaluation of aortic morphology and elastic properties in children and adolescents with Ullrich-Turner syndrome (UTS): High prevalence of aortic changes already in early life**

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**Objective:** Patients with UTS have an increased risk for progressive aortic root dilatation and dissection. Abnormal biophysical wall properties may be responsible for this. To avoid major cardiovascular events, it is important to recognize the high-risk patients early. Aim of this study was to evaluate morphology and elastic properties of the aorta for the first time only in children and adolescents with UTS.

**Methods:** In a prospective, cross-sectional study we examined 37 conscious girls with UTS without cardiac history, and all treated with growth hormone (median age (years): 13.5 [6.7-20.5], median height SDS -2.21 [-3.44- -0.39]), and 12 healthy controls (14 [9-18]) using a 1.5 Tesla whole-body MR scanner. Contrast-free 3D-MR angiographies were performed to assess aortic morphology. Diameters were measured at ascending (AAO) and descending aorta (DAO) and their ratio was calculated. BSA-corrected AAO-diameter is defined as aortic size index (ASI). MR protocol included 2D-CINE MRI to assess the relative change in aortic cross-sectional areas at the level of diaphragm to calculate aortic compliance (C). ECHO and ECG were performed routinely.

**Results:** Changes of aortic morphology were found in 32% of the patients, while some had more than one alteration. Dilatation or prolongation was seen in 7 patients, 9 patients had an increased AAO/DAO ratio >1.5 and 8 patients had an increased ASI > 2 cm/m<sup>2</sup>, representing aortic dilatation. Bicuspid aortic valves were found in 5 patients. Compliance was similar in UTS patients and controls: C= 7.02 ± 2.82 vs. 5.54 ± 2.41 [10<sup>-5</sup>\*Pa<sup>-1</sup>\*m<sup>-2</sup>]; t-test: p=0.13. In routine-ECHO, in only 2 patients an AAO dilatation could be detected. 3 patients revealed a prolonged QT interval.

**Conclusion:** One third of the UTS patients had pathologic aortic findings including the aortic valve, one quarter already exhibits signs of aortic dilatation in early life, although the elastic wall properties seem to be normal yet (under growth hormone therapy). Monitoring of all these parameters may be relevant for evaluating disease progression and treatment options. Therefore, MRI helps to detect morphologic changes early and is more sensitive than ECHO but needs its validation in routine care to improve cardiac morbidity.