Routine Cardiac MRI Screening Reveals Reduced Prevalence of Structural Cardiovascular Abnormalities in Patients With Turner Syndrome

Agnes Hamilton-Baillie, 1,2 Christopher M Jones, 1,2 Caroline Packer, 1,2 Andy Toogood, 2
1 College of Medical and Dental Sciences, University of Birmingham, UK; 2 University Hospitals Birmingham NHS Foundation Trust, Birmingham, UK

Background Turner syndrome has been associated with significantly higher rates of cardiovascular anomalies. Current guidelines for the management of this condition suggest evaluation of the aorta via cardiac magnetic resonance (CMR) imaging or echocardiography every 5 to 10 years to evaluate each patient’s potential for aortic dissection, without prior risk stratification. The current spectrum and frequency of structural cardiovascular anomalies in Turner syndrome is however unclear, with much of the current literature based on historical cohorts.

Purpose To determine through CMR screening the current prevalence of structural cardiovascular anomalies amongst patients with Turner syndrome.

Methods We retrospectively reviewed the medical records of adult patients diagnosed with Turner syndrome who had undergone routine surveillance for structural cardiovascular abnormalities. Each underwent CMR imaging. Data relating to patient demographics and baseline observations were extracted. The presence of structural cardiac anomalies was determined through analysis of MRI and echocardiogram imaging, in addition to review of records relating to previous operative intervention.

Results Medical records for 46 patients with Turner syndrome were reviewed, 7 (15.2%) of whom were mosaic for X-chromosomal monosomy. Median age was 28 years (range 21–73 years). 19/46 (41.3%) patients had no reported structural cardiovascular anomaly. Of the 27 patients with structural anomalies, 19 (70.4%) were reported to feature a bicuspid aorta, 13 (28.3%) coarctation of the aorta and 10 (21.7%) dilated aortic root. 13/46 (28.3%) patients underwent surgical intervention, including 11 (23.9%) who underwent repair of aortic coarctation.

Conclusions The presence of cardiovascular abnormalities amongst our population was lower than that reported within the literature. Patients with Turner syndrome are nevertheless likely to undergo surgical intervention. Risk stratification tools are therefore required to optimise use of resources in patients with Turner syndrome requiring routine screening for structural cardiac anomalies.

Conflicts of interest The authors declare that they have no conflicts of interest relating to this article.

Abstract 16 Figure 1 MAPSE measurements were taken in the four-chamber cine.