International Journal of Cardiology xxx (xxxx) xxx

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Letter to the Editor

Preventing aortic dissection in Turner syndrome: Who faces the risk?[1]

ARTICLE INFO

Keywords
Turner syndrome
Aortic dissection
Prophylactic surgery

As a group that participated in the creation of the international Turner syndrome guidelines [1], we are concerned that the article by Thunstrom et al. "*Aortic size predicts aortic dissection in Turner syndrome – A 25-year prospective cohort study*" [2] reports a high (~ 80%) false-positive dissection rate. The article proposes small echo-determined aortic diameter surgical thresholds to prevent aortic dissection: an absolute ascending aortic diameter of 3.3 cm or Turner syndrome-specific *Z*-score of 2.1 (normal<2) [3]. The international guidelines endorse larger ascending aorta thresholds for surgery: > 4 cm and TS-specific Z-scores ≥4. The consequences of assigning high-risk to individuals with near normal aortic diameters are significant because they would expose numerous people (~ 80%) who are unlikely to dissect to major thoracic surgery, unnecessary physical activity restrictions, and fear of attempting pregnancy. The reason Thunstrom's results differ markedly from prior studies may be that only 4 of 12 echocardiograms were performed within one year before dissection. In the others, aneurysm growth may have accelerated during the intervening years. Also, the international guideline specifies ascending aorta interrogation at the level of the right pulmonary artery to insure consistency. Thunstrom states only that a "proximal portion" of the ascending aorta was measured. Variable diameters along the length of the proximal ascending aorta may have lowered the mean value. The international guidelines are useful but have limitations as they are based on expert opinion, meta-analyses, and registry data. Studies employing CT/MRI imaging and consideration of co-morbidities (age, BAV, coarctation, blood pressure) are necessary [4].

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Michael Silberbach[a,*], Alan C. Braverman[b], Siddharth K. Prakash[c], Jolien W. Roos-Hesselink[d], Emilio Quezada[e], Cindy Scurlock[f]
[a] *Doernbecher Children's Hospital Oregon Health & Sciences University, Portland, OR, USA*
[b] *Cardiovascular Division, Department of Medicine, Washington University School of Medicine, St Louis, MO, USA*
[c] *Department of Internal Medicine, McGovern Medical School, University of Texas Health Science Center at Houston, Houston, TX, USA*
[d] *Department of Cardiology, Erasmus Medical Center, Rotterdam, the Netherlands*
[e] *Department of Pediatrics, University of California San Francisco, CA, USA*
[f] *Turner Syndrome Society of the United States, Houston, TX, USA*

[*] Corresponding author at: CDRC-P, 707 SW Gaines Rd, Portland, OR 97239-3098, USA.
E-mail address: silberbm@ohsu.edu (M. Silberbach).

[1] All authors take responsibility for all aspects of the reliability and freedom from bias of the data presented and their discussed interpretation.

https://doi.org/10.1016/j.ijcard.2023.01.075
Received 12 January 2023; Accepted 25 January 2023
Available online 30 January 2023