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Diagnostic Divergence Based on Z-Score Models in the Evaluation of Patients with Connective Tissue Disorders

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Keywords

Aortopathy, bicuspid aortic valve (BAV), connective tissue disorders, echocardiography, z-score

Abstract

Background

Z-score models in pediatric echocardiographic studies are pivotal in medical decision making in patients with concern for aortopathies. An aortic root Z score > 2 may prompt genetic testing or a clinical diagnosis of Marfan if Z >3. This study sought to determine if there was a significant difference between Boston (BTN) and Pediatric Heart Network (PHN) allometric Z-score models in patients referred for aortopathy with or without connective tissue disorders (CTD).

Methods

We conducted a retrospective analysis of 397 patients for possible aortopathy with or without connective tissue disorders. Study dates: 1/2019-12/2022. Patient sex, age, aortic root (ART), aortic annulus diameter, ascending aortic (AAO) diameter were measured and evaluated with both PHN and BTN Z-score models. Differences between Z-score models were analyzed for each aortic segment in the sample populations. Z ≤ 2 was considered normal.

Results

ART Z-scores had a high degree of correlation between the two models (Pearsons 0.999 (0.999,0.999), Lin's 0.996 (0.962, 0.971)); however, the models diverged at higher Z-scores. Median PHN Z-scores were larger than Boston Z-scores for each aortic segment in both populations but most notable in CTD patients where ART Boston Z = 0.29 (-0.44, 1.39) versus PHN Z = 0.52 (-0.34, 1.82); AAO Boston Z =0.11 (-0.46, 0.77) versus PHN Z = 0.49 (0.42, 0.55). Using PHN, there were 21 additional cases of ART dilation and 2 additional cases of

AAO dilation. Further analysis of ART Z-score measurements in both patient sub-populations demonstrated exaggerated differences between PHN and Boston of + 0.59 Z-scores (0.37, 0.75) in patients with CTD compared to 0.18 (0.10, 0.35) in those without CTD.

Conclusion

The use of PHN versus Boston Z-scores are highly congruent though PHN Z-scores may lead to an increased diagnosis of aortic dilation in patients being evaluated for connective tissue disease.