



Diagnostic value of the
'soccer ball' morphology in
the Marfan aortic root.

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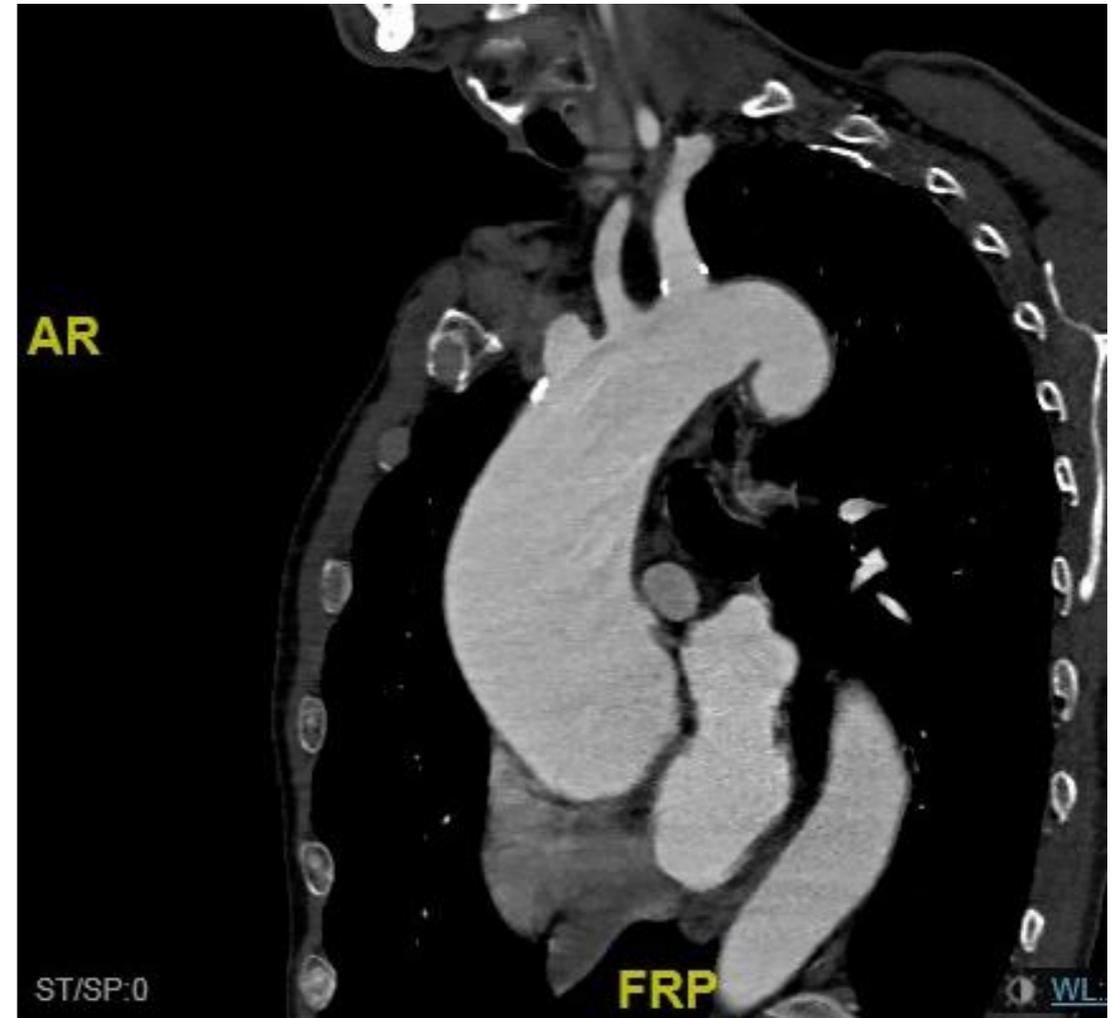
Background

- The diagnosis of Marfan syndrome remains challenging, with aortic diameter featuring heavily in the Ghent nosology.
- Anecdotally aneurysmal Marfan roots are noted to have a typical shape - often described in the literature as 'tulip' or 'onion' bulb.

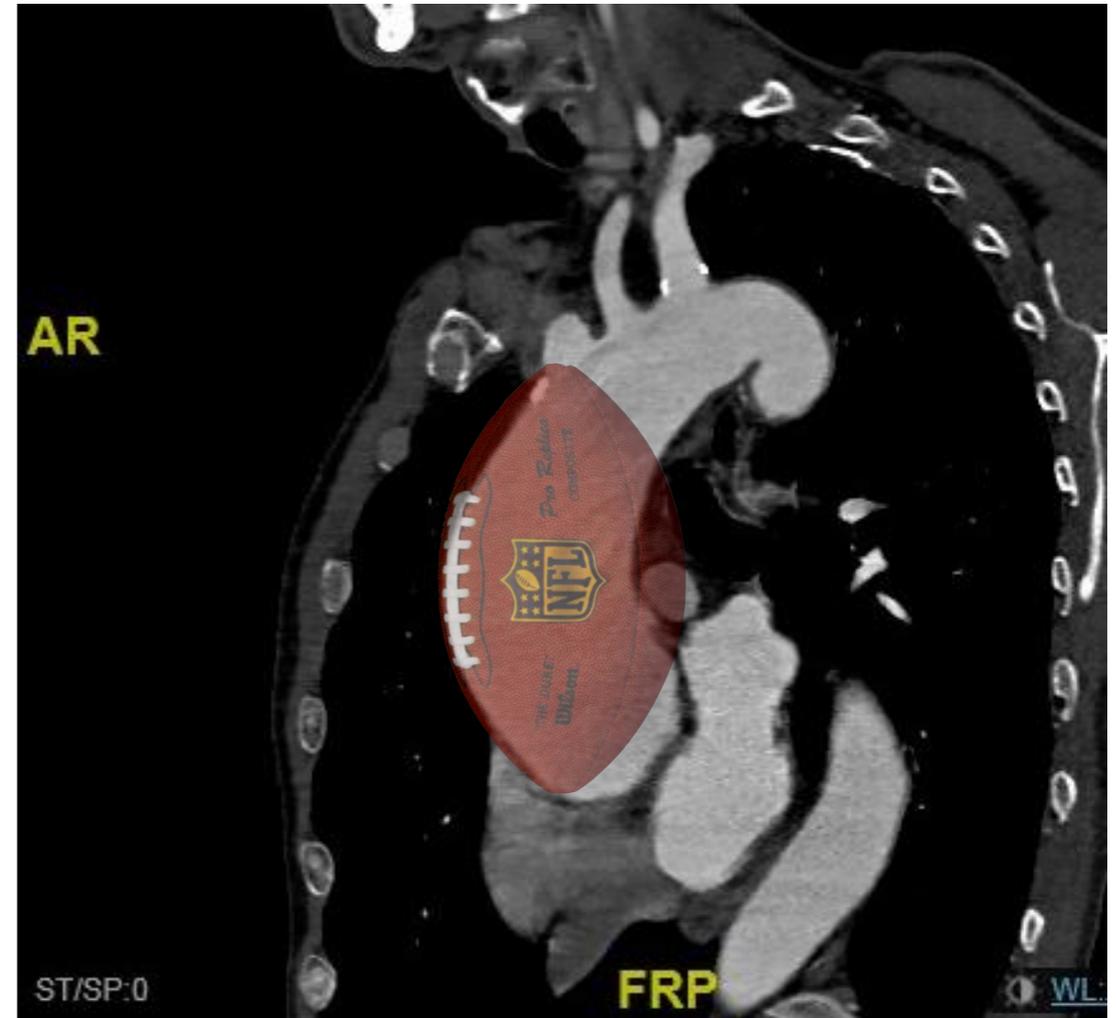
Background

- Similar observations of a characteristic shape made at our institution, and noted in operative documentation.
- No value to colleagues in pathology who receive deflated and/or fragmented tissue.
- Preference given to classifying as 'soccer ball' vs 'rugby ball' / 'other shape' to simplify understanding.

Background Aortic Shape



Background Aortic Shape



Purpose

- We sought to evaluate the predictive accuracy of this morphologic description, and its potential to contribute to the diagnostic panel for Marfan syndrome.

Methods

- All patients who underwent aortic root replacement (ARR) at our institution between 2012 and 2016 were identified from a prospective cardiac surgical database.
- Patients with a diagnosis of Marfan syndrome were identified through electronic case records.
- A single rater, blinded to aetiology, classified the aortic root shape on imaging into 'soccer' or 'rugby'/'other'.
- Descriptive statistics and predictive analytics performed using the Analyse-it® package for Microsoft Excel 365.

Results Demographics

- 97 procedures
 - 74 (76%) elective
- 66 (68%) male
- age 59 ± 15 years
- 9 (9%) Marfan syndrome
- complete data available for 81 (84%)

Results Morphology vs Aetiology

	Marfan Syndrome	Other
Soccer Ball	2 TRUE POSITIVE	6 FALSE POSITIVE
Rugby Ball / Other	6 FALSE NEGATIVE	62 TRUE NEGATIVE

Results Predictive Validity

- Sensitivity = 25% (3 to 65%)
- Specificity = 91% (82 to 97%)
- Predictive Accuracy = 84% (74 to 92%)

Limitations

- single-rater (in feasibility stage)
- no intra-operative photography
- incomplete imaging & diagnostic data

Conclusions

- The 'soccer ball' aortic root morphology showed a low sensitivity but a high specificity for Marfan syndrome.
- This discriminatory feature deserves further evaluation with consideration to inclusion in the Ghent nosology.

Next Steps

- Expansion of retrospective inclusion - 1996 to 2016 - with retrieval of archive imaging.
- Blinded panel of independent raters, from novice (medical student) to expert (consultant).

Thank you.

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