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

<table>
<thead>
<tr>
<th></th>
<th>Marfan Syndrome</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Soccer Ball</strong></td>
<td>2 TRUE POSITIVE</td>
<td>6 FALSE POSITIVE</td>
</tr>
<tr>
<td><strong>Rugby Ball / Other</strong></td>
<td>6 FALSE NEGATIVE</td>
<td>62 TRUE NEGATIVE</td>
</tr>
</tbody>
</table>
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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attendance and this presentation supported by: