Dilated Mid Ascending Aorta in Hypertrophic Cardiomyopathy is Associated with Dynamic Left Ventricular Outflow Tract Obstruction

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**PROBLEM**

- Prior studies have shown that Hypertrophic Cardiomyopathy is associated with Dilated Aorta.
- However, whether this dilatation is due to post-stenotic dilatation from dynamic left ventricular tract (LVOT) obstruction or yet undefined genetic abnormalities has not been clearly demarcated.
- As HCM is an inherited disease, we hypothesized that DA in HCM will be associated more strongly with cardiac sarcomere genetic abnormalities as compared to hemodynamics of dynamic LVOT obstruction.

**BACKGROUND**

- A prior study conducted by our group has shown that Hypertrophic Cardiomyopathy (HCM) is associated with dilated aorta (DA).
- In the same study, out of 201 patients, 18 patients had DA and 13 out of those 18 patients had a higher prevalence of dynamic LVOT obstruction as compared to those HCM patients without DA (72 vs 52%, p=0.1).
- 12 patients with DA were also tested for sarcomere gene mutation, 4 of which tested positive (33.3%).
- In our study, we further analyzed 175 patients, by adding more information to the database, including measuring gradients (resting and induced, n=123), gene testing (n=124), and measuring mid ascending aorta and Sinus of Valsalva in all 175 patients.

**OBJECTIVE**

- To determine whether this aortic dilatation is due to post-stenotic dilatation from dynamic LVOT obstruction or yet undefined genetic abnormalities.

**METHODS**

**Study Population**

- We retrospectively reviewed the medical and echocardiography records of the 175 patients with HCM seen and characterized by AJT in a tertiary-care HCM center.
- Of these, 124 received genetic testing.
- The patients (n=175) were categorized to have significant LVOT obstruction if the baseline dynamic LVOT gradient was >20 mmHg.
- All the patients underwent measurement of the sinuses of Valsalva (SV) and mid ascending aorta (mAA) with leading-edge-to-leading edge technique in diastole.
- The aorta was defined as dilated if it was >4 cm in the SV or mAA.

**RESULTS**

- Out of the 124 patients tested, 56 (45%) were found to be gene-positive.
- Out of all 175 patients, the mean LVOT gradient was 24±34 and a range of 0-160 mmHg, with 49 patients having a gradient >20 mmHg.
- The gene-negative patients had a higher mean dilated SV (3.39 cm vs 3.12 cm; P=0.0038) and dilated mAA (3.3 cm vs 3 cm; P=0.005) than gene-positive patients (n=56).
- Gene-positive patients had a slightly lower prevalence of dilated SV (11% vs. 15%) and mAA (7% vs. 10%), which was not statistically significant.
- With a patients baseline LVOT gradient >20 mmHg had a 3 times higher prevalence (15.1% vs. 4.9%) of dilated mAA (>4 cm) than those with LVOT gradient of <20 mmHg (OR: 3.41, 95% CI 1.12-10.88, P=0.03), whereas no significant relationship was seen with dilated SV (OR: 1.37, 95% CI 0.51-3.43, P=0.5).
- This association with dilated mAA persisted after adjusting for hypertension, aortic stenosis, aortic regurgitation and aortic prosthesis in stratified and multivariate analyses.

**Logistic Regression Analysis for Increased Odds of Dilated mAA with Adjustment for risk factors of dilated mAA**

<table>
<thead>
<tr>
<th>Odds Ratio</th>
<th>95% CI</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baseline mAA</td>
<td>LVOT gradient &gt;20mmHg</td>
<td>9.41</td>
</tr>
<tr>
<td>Adjusted Model 1</td>
<td>LVOT gradient &gt;20mmHg Age</td>
<td>5.79</td>
</tr>
<tr>
<td>Adjusted Model 2</td>
<td>LVOT gradient &gt;20mmHg Systolic BP</td>
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<td>Adjusted Model 3</td>
<td>LVOT gradient &gt;20mmHg Heart Rate</td>
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<tr>
<td>Adjusted Model 4</td>
<td>LVOT gradient &gt;20mmHg Diabetes</td>
<td>4.71</td>
</tr>
</tbody>
</table>

**CONCLUSION**

- The dilatation of mAA in patients with HCM appears to be more strongly associated with baseline dynamic LVOT obstruction than with genetic abnormalities.

**REFERENCES**