Prognostics and Management of Patients with Moyamoya Syndrome and Sickle Cell Disease: a retrospective cohort study

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Disclosures

• None
Moyamoya syndrome, a progressive, idiopathic stenosis of the internal carotid arteries, results in increased risk for both ischemic and hemorrhagic stroke, particularly in sickle cell disease-induced moyamoya.

Revascularization procedures have been shown in small studies to be both safe and efficacious; however, transfusions remain the standard of care and risk factors for recurrent stroke while on transfusions are unknown.
Methods

• Retrospective cohort study including all patients across the Emory University Hospital system who were diagnosed with MMS and SCD
• Patients identified using CPT codes related to their diagnosis of MMS from neurosurgery, neurology, hematology, and radiology records during the catchment window of the years 2015-2019
• Full chart review performed to establish sufficient electronic documentation for data abstraction defined as documentation three years pre- and post-diagnosis with MMS
Figure: Follow up from initial presentation of patients undergoing revascularization. Red square indicates event while on optimum chronic transfusion therapy.
Figure: Follow up from initial presentation of patients maintained on chronic transfusion therapy. Red square indicates event while on optimum chronic transfusion therapy.
<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Regression Coefficients (B)</th>
<th>Significance (P)</th>
<th>Odds Ratio</th>
<th>95% CI (Odds Ratio) Lower</th>
<th>95% CI (Odds Ratio) Upper</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td>0.424</td>
<td>0.600</td>
<td>1.528</td>
<td>0.314</td>
<td>7.437</td>
</tr>
<tr>
<td>Age at First Stroke</td>
<td>-0.116</td>
<td>0.067</td>
<td>0.890</td>
<td>0.786</td>
<td>1.008</td>
</tr>
<tr>
<td>Age at Diagnosis</td>
<td>-0.041</td>
<td>0.386</td>
<td>0.960</td>
<td>0.874</td>
<td>1.053</td>
</tr>
<tr>
<td>Pre-diagnosis number of strokes</td>
<td>2.157</td>
<td>0.043</td>
<td>8.645</td>
<td>1.071</td>
<td>69.768</td>
</tr>
<tr>
<td>Post-diagnosis number of strokes</td>
<td>1.581</td>
<td>0.022</td>
<td>4.858</td>
<td>1.257</td>
<td>18.778</td>
</tr>
<tr>
<td>Total number of previous strokes</td>
<td>2.830</td>
<td>0.008</td>
<td>16.944</td>
<td>2.065</td>
<td>139.027</td>
</tr>
</tbody>
</table>
Results

Survival Functions

Cum Survival

0.0 0.2 0.4 0.6 0.8 1.0

yrsdxtodeath

0.00 5.00 10.00 15.00 20.00

Time Diagnosis to Death (years)
• Significant risk factors for recurrent stroke while on chronic transfusion therapy including total number of strokes, number of strokes before diagnosis, and number of strokes after diagnosis

• The most significant factor with an odds ratio of 16.9 was total number of previous strokes

• Though not statistically significant, there is decreased stroke rate per patient year and mortality rate in the post-operative group of patients. Interpretation likely limited by sample size
Summary

• Despite the relatively small group affected by these co-diagnoses, the burden of disease is universally high suggesting the need for advocacy around earlier radiographic evaluation to appreciate the extent of the disease in patients known to be at risk with scheduled repeat examinations and earlier discussion of revascularization for individuals who have demonstrated higher rates of stroke in the past.