Current Epidemiology of Symptomatic Pediatric Moyamoya in the United States: A National Database Analysis

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Introduction

• In the United States, the majority of literature on pediatric moyamoya is comprised of small institutional based series, limiting the ability to analyze patterns of demographics and presentation of moyamoya on a national scale.
Methods

• A comprehensive search of the Kids' Inpatient Database (KID) was conducted to capture all patients with primary diagnoses of acute ischemic stroke/transient ischemic attacks (AIS/TIA) and intracerebral hemorrhage (ICH).

• Records were reviewed to identify patients with secondary diagnosis of moyamoya disease (MM) amongst the AIS/TIA and ICH cohorts. We then analyzed the resulting three cohorts: AIS/TIA without MM, AIS/TIA with MM, and ICH with MM.
Ischemic stroke/ TIA

- No moyamoya
  - Ischemic stroke/ TIA without moyamoya
- Moyamoya
  - Ischemic stroke/ TIA with moyamoya

Hemorrhage (ICH, SDH, SAH, EDH)

- Moyamoya
  - Hemorrhage (ICH, SDH, SAH, EDH) with moyamoya

Moyamoya syndrome (DS, SCD)
Results

- From 1997-2016, there were 3854 AIS/TIA patients (6.1% with MM) and 7030 ICH patients (0.5% with MM) found in the KID.
- Amongst moyamoya cohorts, 86.4% presented with AIS/TIA and 13.6% presented with hemorrhage; with significantly higher prevalence of African-Americans in the two moyamoya cohorts compared to AIS/TIA without moyamoya ($P < 0.001$).
- The incidence of both AIS/TIA and moyamoya was higher in Southern US.
- A concurrent medical disease was present in 48% of the AIS/TIA with MM (specifically Sickle cell disease (SCD), 28%); which was significantly higher than the other cohorts.
Sex within moyamoya population compared to non moyamoya stroke

P=0.046
Race within moyamoya population compared to non moyamoya stroke

<table>
<thead>
<tr>
<th>Race</th>
<th>Percent moyamoya among stroke population</th>
</tr>
</thead>
<tbody>
<tr>
<td>White</td>
<td>4.98</td>
</tr>
<tr>
<td>Black</td>
<td>10.18</td>
</tr>
<tr>
<td>Hispanic</td>
<td>6.90</td>
</tr>
<tr>
<td>*Asian/Pacific Islander</td>
<td>5.63</td>
</tr>
<tr>
<td>Other</td>
<td>11.21</td>
</tr>
</tbody>
</table>

P = 0.004
Discussion

• Pediatric stroke patients with MM in the US present with AIS/TIA much more frequently than hemorrhage, especially in younger age group.
• There was no statistically significant gender predominance between the 3 cohorts, which is in contrast to reports from Asian literature.
• Pediatric stroke patients in the US are unique in having a high prevalence of co-morbid medical conditions, particularly SCD, in comparison to populations from other countries around the world.
Summary Points

• Moyamoya syndrome is highly prevalent among moyamoya patients who present with stroke in the United States

• Almost 2/3 of African-Americans (62.5%) with moyamoya had a concurrent diagnosis of SCD.

• Overall, these data provide novel findings about symptomatic pediatric moyamoya patients in the US and can help to inform future guidelines on screening and treatment for this condition.