Moyamoya Disease Epidemiology outside of Asia

In a Nationwide Inpatient Sample (NIS), patients presenting with ischemic stroke were more likely to be 65 years and older ($p < 0.001$); patients presenting with intracranial hemorrhage (ICH) were more likely to be 65 years and older ($p < 0.001$), male ($p = 0.027$), and Asian ($p = 0.007$); those presenting with seizure were more likely to be less than 10 years old ($p = 0.002$) and African American ($p < 0.001$); and those presenting with headache were more likely to be between 10 and 19 years old ($p = 0.008$).

The findings demonstrate that the distinct presentations of Moyamoya are associated with patient age, gender, and race. This is the largest study of its kind and adds to the collective understanding of this rare but life-threatening condition [1)].

Studies from outside of Asia are rare. In Washington state and California, the incidence of MMD was reported to be 0.086/100,000 based on 298 patients. The incidence was the highest in Asians, followed by Blacks, Whites, and Hispanics. The incidence in Asian Americans was 4.6 times higher than that in Whites. Female preponderance was also noted [2)].

African-Americans had an earlier disease onset with a median age of 18. However, a more recent study based on the Nationwide Inpatient Sample database reported that MMD appears to be distributed among the races according to their relative proportions in the USA population [3)].

From 2005 to 2008, there were an estimated 7,473 (2,236 pediatric and 5,237 adult) patients admitted with a diagnosis of MMD in the USA. MMD patients were most frequently Caucasians. Overall, ischemic stroke was the most common reason for admission in both children and adults. Hemorrhagic stroke was more frequent in adults compared with children, and there was a bimodal age distribution with peaks in the first and fourth decades of life. Female-to-male ratio was 2.2. Thus, MMD in the USA does not seem to differ from East Asian MMD.

The incidence of Moyamoya disease (MMD) in Europe is not well known. In those affected, the risk of brain hemorrhage is considered low. A study of Birkeland et al. aimed to investigate the incidence and clinical presentation of MMD in the Danish population.

Eligible patients were identified in the Danish National Patient Register from 1994 to 2017. They collected clinical and radiological data from individual patient records from neurological, neurosurgical, and pediatric units across Denmark. The diagnosis was validated according to established criteria. They also extracted basic demographic data on the cohort from the Danish Civil Registration System.

A total of 52 patients fulfilled the diagnostic criteria for MMD. Most cases were native Danes and only 15% of cases had an East Asian background. The ratio of female to male patients was 1.8, and the incidence had two peaks: one in childhood and another in young middle age. Until 2007, MMD was only diagnosed sporadically. From 2008 onwards, the incidence rate was 0.07 per 100 000 person-years (95% confidence interval 0.05-0.09 per 100 000 person-years). The most common mode of

presentation was ischemic stroke (33%), followed by hemorrhage (23%), headache (17%), and transient ischemic attack (14%).

MMD is rare in Denmark, but associated with a considerable risk of hemorrhage. Thus, MMD should be considered in the workup for ischemic as well as hemorrhagic stroke in children and middle-aged Caucasians [4].

References

1)
Fuentes AM, Chiu RG, Mehta AI. Disparities in the symptomatic presentation of Moyamoya disease in the United States: A nationwide all-payer analysis. J Clin Neurosci. 2021 May;87:92-96. doi: 10.1016/j.jocn.2021.02.019. Epub 2021 Mar 20. PMID: 33863543.
2)
Uchino K, Johnston SC, Becker KJ, Tirschwell DL. Moyamoya disease in Washington State and California. Neurology. 2005;65(6):956-958. doi:10.1212/01.wnl.0000176066.33797.82
3)
Kainth D, Chaudhry SA, Kainth H, Suri FK, Qureshi AI. Epidemiological and clinical features of moyamoya disease in the USA. Neuroepidemiology. 2013;40(4):282-287. doi:10.1159/000345957
4)
Birkeland P, Tharmabalan V, Lauritsen J, Ganesan V, Bjarkam CR, von Weitzel-Mudersbach P. Moyamoya disease in a european setting: A danish population-based study [published online ahead of print, 2020 Jul 15]. Eur J Neurol. 2020;10.1111/ene.14439. doi:10.1111/ene.14439