Response to Letter Regarding Article, “Perfusion Characteristics of Moyamoya Disease: An Anatomically and Clinically Oriented Analysis and Comparison”

We thank Mugikura and Takahashi¹ for their thoughtful analysis of our data on cerebral perfusion characteristics in Moyamoya disease (MMD).² The authors present a detailed discussion of particular compensatory mechanisms provided by the posterior cerebral artery (PCA), namely a descriptive subcategorization into deep (proximal PCA to pericallosal artery) and more superficial collateralization (distal PCA to middle cerebral artery and anterior/posterior watershed). On the basis of our findings of relative preservation within the pericallosal territory, the authors hypothesize an insufficiency predominantly of superficial collateralization pathways. This particular territory may even be more anteriorly, if the PCA itself features proximal steno-occlusive changes. Indeed the authors bring to our attention an important, previously attested interrelation of PCA involvement in MMD and cortical watershed infarction, which initially may be pronounced more anteriorly, whereas disease progression can also lead to infarction of posterior watershed territory.³

Although spontaneous collateralization is the characteristic hallmark of MMD, it is essential to note that its angiographic evidence alone may not suffice to determine adequacy of compensation or disease severity. It is generally agreed on that intracranial (deep or superficial) compensatory mechanisms are important factors both for grading of disease severity and for prognostic implications,⁴ and the relevance and specific contributions provided by defined steno-occlusive lesions (be it within the PCA or elsewhere) would benefit from further investigation. However, additional spontaneous extra- to intracranial collateralization is another typical feature frequently observed with variable symptomatology.

In this respect, type and localization of collateralization may only help to appreciate the complexity of the predominant mechanisms evolved, whereas the combination of clinical symptoms, angiographic findings, and perfusion measurements are required to assess overall disease burden adequately.

Moreover, it is our impression that the existence of likely distinct subentities of MMD—such as pediatric and adult onset MMD, Asian and European/North American MMD, with or without PCA involvement—is suggestive of an even more heterogeneous pathophysiology with variable expression and progression, rendering a broad generalization even more difficult and vulnerable to inadequacy.

Because some patients do not feature PCA involvement with proximal stenosis or occlusion, we limited our analysis to the most frequent and uniform presentation of MMD (ie, patients with a characteristic anterior rete mirabilis compensating for a progressive stenosis of the internal carotid artery or middle cerebral artery). We are hopeful that our data will help to provide a general understanding of collateralization potential and actual perfusion, but the comment by Mugikura and Takahashi¹ fittingly illustrates the need for an even more detailed approach to this particularly complex disease.

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Disclosures

None.

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