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Guest Editorial

Moyamoya: past, present, future

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This special issue of the Journal of Pediatric Neuroradiology is devoted to moyamoya disease and syndrome. Extensively researched, yet poorly understood, moyamoya literally means, "feeling confused, curious, foggy". Although the classic notion of "puff of smoke" is well accepted in the scientific community, the term moyamoya has a different connotation to a Japanese medical student: "When I am talking to someone and I don't really fully understand, I would say that I am moyamoya." In this sense, moyamoya perfectly describes this ill-defined progressive cerebral vasculopathy, which can be unilateral or bilateral, and involve the anterior or posterior circulations. Nearly 60 years after its initial description, our understanding of the etiology and underlying mechanisms leading to moyamoya remains elusive. For example, the fundamental risk factors and predictors for disease development and progression or the optimal timing of surgical intervention are still unclear. The purpose of this special issue was to engage physicians and scientists from various disciplines to share their current understanding and management approach for children with moyamoya, and discuss future neurosurgical and endovascular therapies whose efficacies can be assessed objectively by neuronal biomarkers and advanced imaging techniques.

This multidisciplinary effort involves specialists from pediatric neuroradiology, neurology, hematology, neurosurgery, and interventional neuroradiology. Bosemani et al. review the imaging findings and differential diagnosis in moyamoya disease and syndrome, highlighting the critical role of neuroimaging in moyamoya management. Advanced imaging techniques are described by Choudri et al, who present a multimodality approach for the assessment of anatomic and perfusion variables in moyamoya, and explain how this information aids in treatment planning and patient monitoring. The authors also discuss the diagnostic and management challenges presented by asymptomatic patients and emphasize the importance of interpreting the results of physiologic imaging studies in a multidisciplinary forum including neurologists and vascular neurosurgeons.

Carpenter et al, review the etiology and clinical manifestations of moyamoya and provide an insight into a specific subset of patients at high risk for the development of moyamoya, i.e., children with sickle cell disease. Medical therapies, including steroids, vasodilators, anticoagulants, calcium channel blockers, and aspirin, have been tried with variable success, yet the only currently effective therapy involves surgical revascularization. The surgical options for moyamoya are reviewed by Rhee et al, who discuss the relative advantages and disadvantages of direct versus indirect bypass procedures, and underscore the importance of maintaining specific physiologic parameters to prevent peri-operative ischemia. Although surgical revascularization has been shown to be beneficial, the timing of intervention in asymptomatic patients and in those with borderline imaging features requires further investigation.

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Schneider et al., emphasizes the critical role of diagnostic cerebral angiography and reviews the currently available endovascular treatment options for moyamoya. As described by Watkins and Heran [1] in a previous issue of the Journal of Pediatric Neuroradiology, catheter angiography remains the gold standard diagnostic modality for neurovascular pathologies, in this case moyamoya. Despite the increased utilization of non-invasive imaging, selective vessel catheterization provides unparalleled information that is crucial for surgical treatment planning. Special attention to radiation dose reduction, by utilizing pediatric low dose protocols and removing radiation scatter grids when possible, while maintaining physiologic parameters as discussed by Rhee et al., allow for cerebral angiography to be performed with minimal risk in this patient population.

This issue ends with a rare example of collateral circulation in a severe case of moyamoya syndrome, in which the vasocorona, the pial arterial network that loosely interconnects the anterior and posterior spinal arteries, acts as a previously undescribed collateral pathway for the posterior fossa arterial vascularization. This vascular configuration is beautifully illustrated by Lydia Gregg, BS, CMI, who also designed this special issue's cover art.

The study of moyamoya disease and syndrome provides an opportunity for collaboration amongst distinct subspecialties, striving toward patient centered, disease specific care. I thank all the authors for their contributions and urge our readers to embrace the multidisciplinary approach highlighted in this issue to address the current and future areas of research aimed at improving the diagnosis and optimal management of patients with moyamoya. These combined efforts will lead to a more comprehensive understanding of the etiology, pathophysiology, potential role for neuronal biomarkers, and more optimally timed and efficacious interventions. With a greater appreciation of the disease process and mechanisms, starting at the cellular level, the haze and confusion that currently characterizes moyamoya will hopefully recede in favor of a future in which we will ultimately be able to alter the natural history of this progressive vasculopathy.

References

[1] Watkins TW, Heran MKS. The role of digital subtraction cerebral angiography and the Wada test in the pediatric population. J Pediatr Neuroradiol 2013;2(3):393–400.