Jiro Suzuki

Moyamoya Disease

With 145 Figures

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Professor JIRO SUZUKI, M. D.

Tohoku University School of Medicine
Institute of Brain Diseases
Division of Neurosurgery
2-1 Seiryo-Cho, Sendai 980, Japan

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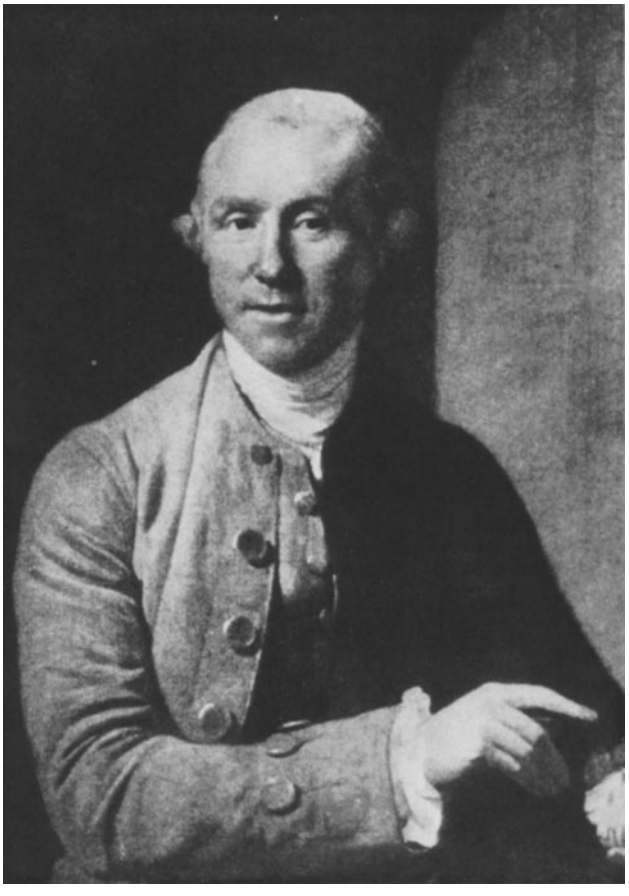

JOHN HUNTER (1728–1793)

"Blood goes where it is needed," he said in 1785

Moyamoya vessels are collateral pathways of the brain
JIRO SUZUKI (1963)

Preface

The first mention of moyamoya disease as a distinct disease entity was in a paper I published in 1965. The abnormal net-like vessels at the base of the brain seen in cerebral angiograms of this disease were described by most native speakers of Japanese as "moyamoya," a Japanese expression for something hazy, such as a puff of cigarette smoke drifting in the air. In fact, prior to my advocacy of this term, this type of vascular network was often referred to as "moyamoya vessels" by Japanese researchers.

In 1969, Dr. A. Takaku and I submitted a paper to the *Archives of Neurology* entitled "A Disease Showing Abnormal Net-like Vessels at the base of the Brain," with a subtitle of "Moyamoya Disease." The editor, however, interchanged the main title and the subtitle and brought this term "moyamoya" to fame! Since then, researchers throughout the world have been using this poetic word! For a nicknaming godfather like me, it is a special joy to have this Japanese word enter the honored ranks of medical terminology.

My first publication concerning this lesion was a report discussing six cases in 1963. Therefore it is now 20 years since I first became aware of and started to study moyamoya disease. At first, I merely thought that it was a strange vascular network, unique to these first few patients, and I reported these cases as being cases of acquired collateral vessels. Thereafter, many researchers speculated that it might be a form of vascular tumor, and intense debate as to whether this disease is congenital or acquired began. Currently, it is generally thought that moyamoya vessels are collateral pathways formed due to gradually progressing stenosis of the terminal portion of the internal carotid artery, for reasons unknown.

Although the incidence of moyamoya disease is still said to be higher in Japan than elsewhere, in recent years there have been reports of cases from both the East and the West, and moyamoya disease has become a matter of great interest to researchers throughout the world.

Through the combined efforts of many researchers over the past 2 decades, many aspects of moyamoya disease have been clarified. These include the incidence according to age and sex, symptomatology, angiographical characteristics, electroencephalographical characteristics, and details of the cerebral blood flow in such cases. This is not to say, however, that unresolved problems no longer exist. Most importantly, it is still unclear what causal factors are involved in the progressive narrowing and occlusion of the terminal portion of the internal carotid artery bilaterally. Since the vascular abnormalities at this portion are the essence of the disease, it is apparent that some fundamental questions still need to be answered.

Nevertheless, as readers of this volume will understand, our systemic autopsy studies and animal research have suggested that moyamoya disease may be caused by immunological arteritis throughout the body, which manifests itself most easily and most severely at those cerebral arteries which receive innervation from the superior cervical sympathetic ganglia.

Having actively researched this disease for 20 years, we, the Sendai Group of Neurosurgeons, decided to summarize our experiences and research in a single volume. With each of the researchers reporting on his own research interests related to moyamoya disease, we hav tried to cover all aspects of this disease, including its etiology.

I am hopeful that this volume will serve as an introduction and an up-to-date summary of our current knowledge concerning moyamoya disease for physicians and researchers throughout the world.

I would like to express my thanks to all my colleagues and to the nurses and paramedical staff for their untiring efforts during this period. I also acknowledge, with thanks, the valuable assistance of Mr. Norman D. Cook in improving the English throughout the book. Finally, I must say that this publication has been made possible by the enthusiasm and efforts of the various doctors (T. Wada, N. Kodama, T. Kayama, H. Niizuma, H. Ohyama, T. Yoshimoto, A. Ogawa, N. Nakamura, Y. Sakurai, S. Komatsu, S. Kanayama, Y. Nagamine, T. Onuma, N. Kasai, T. Yonemitsu, T. Watanabe, M. Suzuki, R. Katakura and A. Takahashi) in our department, and especially Drs. S. Fujiwara, H. Seki and M. Kameyama.

JIRO SUZUKI

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