# Contents

## Part 2 Indirect Revascularization

# Foreword

I have to admit that during the course of my professional career, I have not encountered anything as fascinating as moyamoya disease (MMD). I can still remember those days when MMD was considered a rare disease, mostly seen in patients from Japan, where it was originally described more than 50 years ago. It was fascinating to see the unusual cerebral angiograms of these patients, showing stenoocclusive changes to the brain-supplying arteries in combination with numerous newly formed small collateral channels at the base of the brain. These changes were difficult to comprehend, particularly when compared with vascular changes noticed in common cerebral ischemia. When it came to treating these patients, we had the option of choosing from a variety of procedures, again mostly introduced by our colleagues from Japan. These procedures were later on summarized under the broad category of "indirect cerebral revascularization." Moreover, it was found more difficult from a technical point of view to perform an extraintracranial arterial bypass using the superficial temporal artery as the donor vessel. This was, however, not because the epicerebral recipient vessels were smaller in diameter in comparison to the situation in chronic cerebral ischemia. It was discovered later on that the cortical arteries in patients with MMD have a different morphologic design, with a thinner structure of the arterial wall, which in turn requires increased attention and a greater amount of skills when performing a direct end-to-side anastomosis.

Initially, we came across these patients only rarely, maybe two or three cases per year. But this has changed drastically over the years. At the end of my career, the number of patients with MMD had increased to about 30 to 40 per year, and it was no longer a local phenomenon. Meanwhile, sizable clinical series of patients with MMD have been published from centers all over the world.

So, what do we know now about MMD that we did not know 25 years ago? The lessons from clinical experience and related research data have further substantiated that MMD is a particular form of ischemic cerebral disease that can be differentiated from more common entities of cerebrovascular occlusive disease beyond characteristic angiographic findings. Based on functional studies, we now know that MMD is representative of hemodynamic cerebrovascular insufficiency. A further distinctive feature of MMD is the unique capability of the brain to create new collateral inflow channels to compensate for the impaired blood flow due to the underlying stenoocclusive process within the basal arteries. This is clearly illustrated in patients with advanced MMD, where angiographic findings demonstrate arterial collaterals from meningeal and even extracranial arteries, which is never observed in common cerebrovascular diseases. Considering this observation, surgical revascularization is the logical treatment of choice in patients with MMD.

In fact, it can be viewed as an enhancement of an underlying and ongoing natural process. Even in the absence of randomized clinical trials, it is now generally accepted that surgical revascularization is the only effective treatment for patients with MMD. This is further supported by clinical information derived from large postoperative follow-up studies.

It should be mentioned that this is good news for the field of vascular neurosurgery in general. It was not long ago that in the larger context of vascular neurosurgery, MMD was only mentioned under the heading "miscellaneous." The situation is significantly different now; with extraintracranial bypass surgery for cerebral ischemia becoming obsolete, patients with cerebral aneurysms are increasingly being treated by interventional means, and patients with cerebral AV malformations are being referred to stereotactic radiosurgery. In view of these developments, it is difficult to provide a young colleague with an interest in vascular neurosurgery with good advice on what to do in the future, and I am happy that I do not have to answer this question for myself. However, I'm convinced that the management of patients with MMD will become more important in the future, especially in view of the fact that this disease is still underdiagnosed. It is also good to know that each patient with MMD is usually a candidate for two surgical procedures. There is obviously great potential for further research activities in relation to MMD.

This would not only include research on epidemiology and genetics of MMD but also on its pathophysiology, based on contemporary techniques of molecular biology and other techniques that have become available more recently. We require further

information on questions such as what is the optimal surgical technique, if there is one, and if we should use different surgical approaches for pediatric and adult patients with MMD.

We also need more long-term follow-up studies involving our operated patients, and I am quite sure that there will be more surprises.

Finally, coming back to my personal fascination with MMD that I mentioned in the beginning, let me give you another example for purposes of illustration.

I found it most intriguing to study postoperative angiograms in patients who underwent a combined revascularization procedure 1 or 2 years earlier.

It was amazing to see the number and size of the muscular arterial branches that had ingrown and found connection with the cortical arterial network!

Sometimes, it is difficult to differentiate these newly formed muscular branches from the original extraintracranial arterial bypass. This is another unique feature of MMD, and one wonders if the identification of this mechanism or the isolation of the factor that enables this ingrowth of vessels into the brain could be used for the treatment of other ischemic brain conditions as well.

I am grateful that my long-term coworker Peter Vajkoczy has obviously inherited this interest in MMD, and I will follow his future work with great interest.

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# Preface

Moyamoya vasculopathy (MMV) is a rare cerebrovascular disease that is characterized by bilateral progressive steno-occlusion of basal cerebral arteries, with the emergence of coexisting abnormal net-like vessels. In moyamoya disease, MMV is the single manifestation, whereas in moyamoya syndrome or quasi-moyamoya, MMV is associated with a potentially underlying disease such as a genetic disorder or other coexisting pathology. Although MMV is most frequent in Asian countries, it is ranked among the most frequent causes of stroke in children and adults across the world. The incidence of MMV is on the rise due to increasing awareness of the disease.

The relevance of surgical treatment of moyamoya disease by way of bypass revascularization is undisputed, which is in contrast to the surgical treatment of atherosclerotic carotid artery occlusion. The main aims of revascularization are to restore the blood supply to stabilize cerebrovascular hemodynamics and to regress the fragile moyamoya vessels in order to prevent bleeding. A successful improvement or normalization of cerebral hemodynamics will then result in secondary stroke prevention and improved neurological or neurocognitive outcome. Consequently, bypass surgery for MMV has become an integral part of the clinical practice of many microvascular neurosurgeons around the world. While the role of bypass surgery is well accepted, a versatile range of surgical techniques and strategies exists in the field, which makes it difficult to determine and appreciate the subtle nuances of the varied surgical strategies.

Therefore, it seemed logical to create an instructive manual for neurosurgeons with a step-by-step guide to the surgical techniques. The focus of this book is on introducing neurosurgeons (and other physicians involved in the treatment of these patients) to the different surgical techniques, to the inherent strengths and weaknesses of each technique, and to the surgical considerations that need to be kept in mind. We are grateful to the contributing authors, who are all authorities in their respective fields, for sharing their unique knowledge and expertise with the readers. The descriptions provided by each of them are characterized by an expert assessment of the distinct surgical techniques and their variations, as well as by a standardized illustration of the surgical steps. This book will thus serve as the key manual for everyone interested in the treatment of these complexities and for those who find it a rewarding experience to treat these patients.

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