American Journal of Transplantation 2012; 12: 1353–1355
Wiley Periodicals Inc.

doi: 10.1111/j.1600-6143.2011.03934.x

Letter to the Editor

Organ Donation and Moyamoya Disease

To the Editor:

The Moyamoya Syndrome is a rare cerebrovascular condition characterized by progressive stenosis of the intracranial internal carotid arteries and their proximal branches resulting in the development of severe collateral circulation and associated with a high risk of stroke (1). The Japanese term "moyamoya" was used for the first time in 1969. Suzuki and Takaku reported the presence of abnormally dilated collateral vessels appearing as "a hazy, like a puff of smoke", corresponding to a network of dilated vessels occurring in the region of the circle of Willis (2). Moyamoya disease affects patients from many ethnic backgrounds, including American and European populations, with a higher incidence in persons from Asian heritage. The condition is rare with studies reporting an incidence of 0.086 cases per 100 000 persons in the United States (3).

To our knowledge, there are no reports describing the use of organs from donors diagnosed with Moyamoya disease or studies evaluating the frequency of organ usage from individuals with catastrophic brain injury associated with this disease. Similarly, little is known about the complications associated with the use of solid organs from donors diagnosed with this rare condition. We recently evaluated a patient with massive hemorrhagic stroke who was diagnosed with Moyamoya disease (Figures 1–3). The patient's family agreed to allow organ donation (heart, kidneys, liver and lungs) resulting in successful organ transplantation in five transplant recipients. Evaluation of the patients after a short follow-up period (6 months) confirmed excellent allograft function in all recipients.

We performed a review of the English language literature on the PubMed database using the terms "Moyamoya",

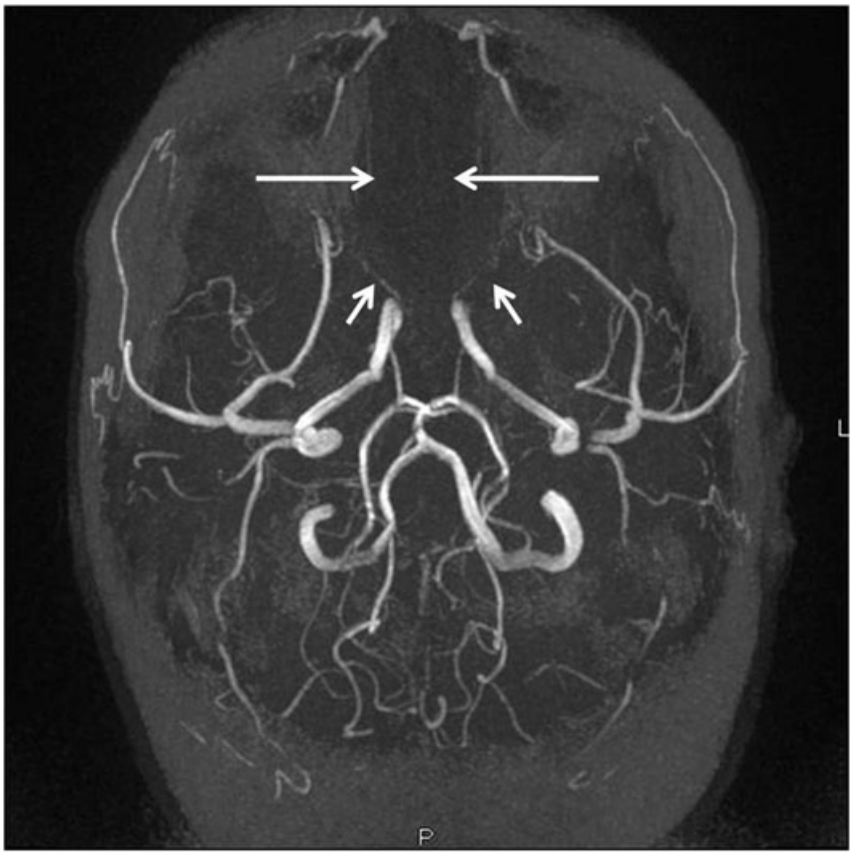

Figure 1: Magnetic resonance angiogram of brain showing areas of severe stenosis in the middle cerebral arteries (short arrows) and anterior cerebral arteries (long arrows).

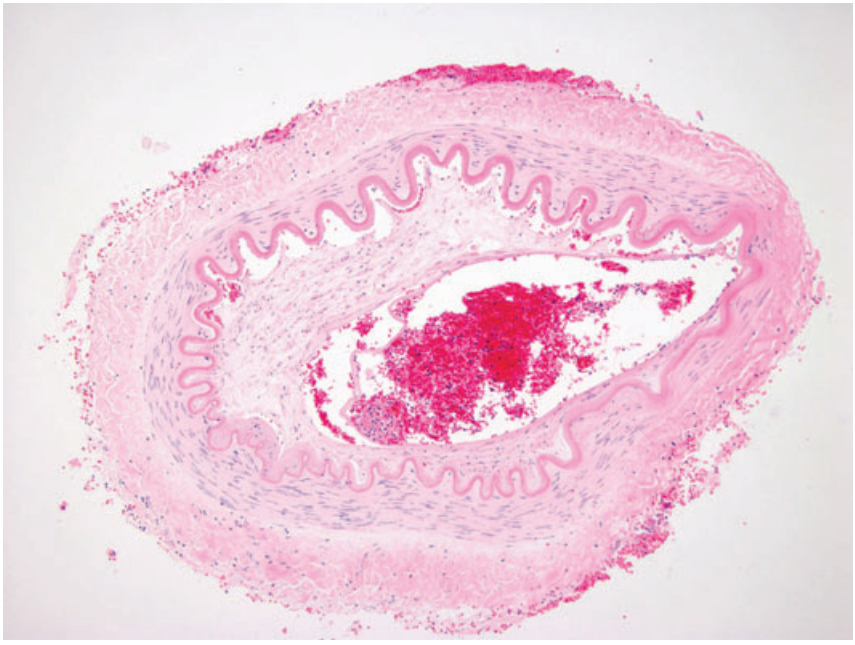

Figure 2: Hematoxylin and eosin-stained section of the right middle cerebral artery, showing fibro–cellular intimal hyperplasia, intact and undulating internal elastic lamina and an attenuated media.

"organ donation" and "stroke" and we were unable to identify reports of successful transplantation using organs from donors diagnosed with Moyamoya disease. In addition, we performed a review of the UNOS database (dates: 1996–2010) and found that Moyamoya disease was reported rarely as cause of death among donors (three thoracic transplants and three abdominal transplants). These findings contrast with reports that suggest a higher number potential number of organ donors with Moyamoya disease and catastrophic brain injury. It is possible that this discrepancy may be explained by coding errors in the UNOS database or misdiagnosis of the disease.

Moyamoya disease is considered a nontransmissible, nonatherosclerotic and noninflammatory condition and, therefore, there are no theoretical risks related to the use of organs from donors diagnosed with this condition. Nevertheless, Moyamoya disease may have extracranial manifestations. Moyamoya has been associated with coronary

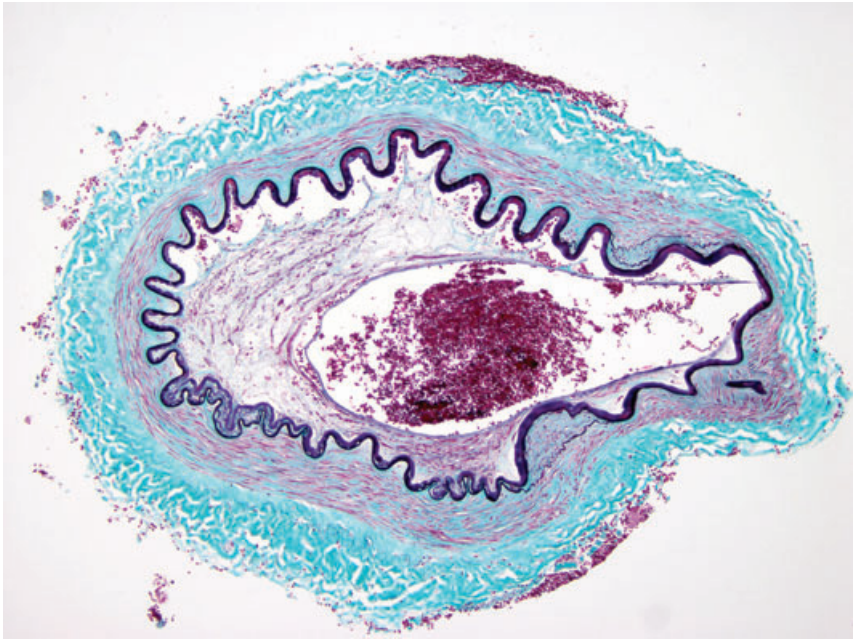

Figure 3: Trichrome-stained section of the right middle cerebral artery showing the intact nature of internal elastic lamina.

artery fistulas and stenosis (4,5) and congenital heart disease (coarctation of the aorta, ventricular septal defect and valve stenosis; Ref. 6), renal artery stenosis and renovascular hypertension (7,8).

We believe that transplant physicians should continue using organs from donors with Moyamoya disease but must be aware of the possible extracranial associations. The long-term complications and outcomes associated with solid organ transplantation from donors diagnosed with Moyamoya disease require further investigation.

Disclosure

The authors of this manuscript have no conflicts of interest to disclose as described by the *American Journal of Transplantation*. There was no funding source needed for this manuscript. The manuscript represents original work that is not being considered or has been accepted for publication elsewhere.

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