


Pial Synangiosis Ameliorates Movement Disorders in the Absence of Prior Stroke in Moyamoya Disease

Journal of Child Neurology
1-6
© The Author(s) 2015
Reprints and permission:
sagepub.com/journalsPermissions.nav
DOI: 10.1177/0883073815609152
jcn.sagepub.com


Stephanie Greene, MD¹, Lalit Bansal, MD², Keith A. Coffman, MD³, Raffaele Nardone, MD⁴, and Giulio Zuccoli, MD⁵

Abstract

Background: Moyamoya disease is a rare cerebrovascular disease characterized by progressive stenosis of the bilateral distal internal carotid arteries and their proximal branches. Both chorea and dystonia have been reported as the initial presentation of moyamoya disease. **Objective:** The objective was to define the clinical presentation and describe the disease course following pial synangiosis of 3 patients with dyskinesias. **Methods:** A retrospective chart review of 3 cases of patients presenting with movement disorders and ultimately diagnosed with moyamoya disease was performed. **Results:** The authors present a case series of 1 patient with dystonia and 2 patients with chorea, all diagnosed with moyamoya disease. All patients experienced resolution of their movement disorders following pial synangiosis. Magnetic resonance imaging disclosed moyamoya disease-related basal ganglia anomalies in all patients. **Conclusions:** Moyamoya disease is an important and surgically treatable cause of movement disorders.

Keywords

moyamoya, chorea, dystonia, movement disorder, pial synangiosis, indirect bypass, pediatric

Received June 15, 2015. Received revised August 4, 2015. Accepted for publication August 22, 2015.

Moyamoya disease is a rare cerebrovascular disease characterized by bilateral stenosis or occlusion of the distal internal carotid arteries and/or their proximal branches. There is compensatory development of a network of dilated leptomeningeal, thalamoperforator, and transdural collateral vessels which give rise to a characteristic angiographic appearance.¹ The incidence of moyamoya disease in the western United States is 0.086 per 100 000,² with a higher incidence reported in Asian countries³ and a female predominance of nearly 2:1.⁴ Onset typically occurs in childhood or early adulthood, and the prognosis in the absence of surgery is 1 of progressive neurologic decline.⁵ Of patients, 20% have severe neurologic impairment or die by adulthood.⁶ The mortality rate in children is 4.3%.⁷ Neurological deficits can arise as a result of ischemia from arterial occlusion or hemorrhage from an ectatic collateral vessel. The most common neurological sequelae are cerebrovascular accidents (transient ischemic attacks, stroke, or hemorrhage), epilepsy, and cognitive decline.^{3,8} Movement disorders associated with moyamoya disease have been reported, with chorea the most frequent.⁹⁻¹¹ To the best of the authors' knowledge, only 23 cases of chorea and 2 case of dystonia s in pediatric patients with moyamoya disease have been described in the English literature.⁹⁻¹⁷ The authors report 3 new cases linking dystonia (1 case) and chorea (2 cases) with

moyamoya disease in children. The literature regarding the incidence, pathophysiology, diagnosis and treatment of these conditions in patients with moyamoya disease is reviewed.

Methods

Institutional Review Board approval was obtained for this retrospective study. The operative database maintained by the department of Neurological Surgery at Children's Hospital of Pittsburgh was queried for patients undergoing indirect bypass for moyamoya between the years of 2009 and 2015. Four patients with movement disorders were identified. One was excluded from further analysis because he

¹ Department of Neurological Surgery, Children's Hospital of Pittsburgh, University of Pittsburgh Medical School, Pittsburgh PA, USA

² Department of Neurology, Children's Hospital of Pittsburgh, University of Pittsburgh Medical School, Pittsburgh PA, USA

³ Department of Neurology, Children's Mercy Hospital, Kansas City, MO, USA

⁴ Department of Neurology, Paracelsus Medical University, Salzburg, Austria

⁵ Department of Radiology, Children's Hospital of Pittsburgh, University of Pittsburgh Medical School, Pittsburgh PA, USA

Corresponding Author:

Stephanie Greene, MD, Children's Hospital of Pittsburgh, 4401 Penn Ave, 4th Fl Faculty Pavilion, Pittsburgh, PA 152, USA.

Email: stephanie.greene@chp.edu

suffered a preoperative stroke at the time of initial presentation, with unsurprising persistence of dystonia after surgery. The 3 remaining patients are being actively seen in neurosurgery clinic at present, and the clinicians making their diagnoses of movement disorder preoperatively were involved in the creation of this manuscript. Patient charts and imaging were reviewed such that patient confidentiality was maintained at all times. By convention, bilateral cases without a disease known to be associated with moyamoya (the radiographic finding) such as Down syndrome or neurofibromatosis are referred to as moyamoya disease, while cases with unilateral stenosis and those with an associated condition are referred to as moyamoya syndrome. Each patient underwent a comprehensive evaluation for his or her movement disorder, and was begun on medication as deemed appropriate. Each patient diagnosed with moyamoya (the radiographic finding) on magnetic resonance imaging (MRI) underwent a cerebral angiogram with isolated external carotid artery catheterizations to assess the candidacy of the superficial temporal arteries for indirect bypass. Each patient underwent an MRI with an acetazolamide perfusion study to assess their cerebrovascular reserve as a marker of stroke risk, and to provide a baseline for postoperative comparison; this noninvasive technique is being published in a separate manuscript by these investigators. Each patient underwent pial synangiosis as described by Scott et al⁸ for treatment of his or her moyamoya disease (or syndrome in patient 4's case). Briefly, the superficial temporal artery was auscultated with Doppler ultrasound and dissected free of the scalp over an 8- to 10-cm length under the microscope. The temporalis muscle was divided, and a sizable craniotomy created. The dura and arachnoid were widely fenestrated, and the superficial temporal artery was sutured to the pia in 4 locations with #8-0 Prolene suture. The dura was left open, the bone replaced, and the temporalis and skin closed. There were no surgical complications, and no postoperative strokes. Imaging, both pre- and postoperatively, and clinical data are described below.

Case Reports

Patient 1 presented in February 2011 as a 16-year-old female with a 2-year history of intermittent right hemcranial headaches and a 4-year history of episodic left face, tongue, hand, and foot dystonic movements. The headaches had remained at their baseline frequency of 4-5 per month, but the dystonic spells had increased in frequency over a period of a few months from 3-5 per month to 2-3 episodes per day. The episodes were characterized by paroxysmal painful dystonic posturing of the face and tongue, clenching of the left hand, and inversion of the left foot. The family history was notable for migraine headaches in her father and 1 sister, and seizures in another sister. There was no family history of dystonia. On physical examination, dystonic tremors of the left foot and ankle could be elicited with sustained isometric gripping of the bilateral upper extremities. These episodes were captured on electroencephalogram, with no electrical correlation. An MRI obtained at an outside institution in the month of the onset of dystonia was of insufficient quality to identify moyamoya. She was started on oxcarbazepine and topiramate 2 months after diagnosis, with complete resolution of her dystonia over the next 2 months. She was referred to a movement disorder specialist at Children's Hospital of Pittsburgh, and a repeat MRI

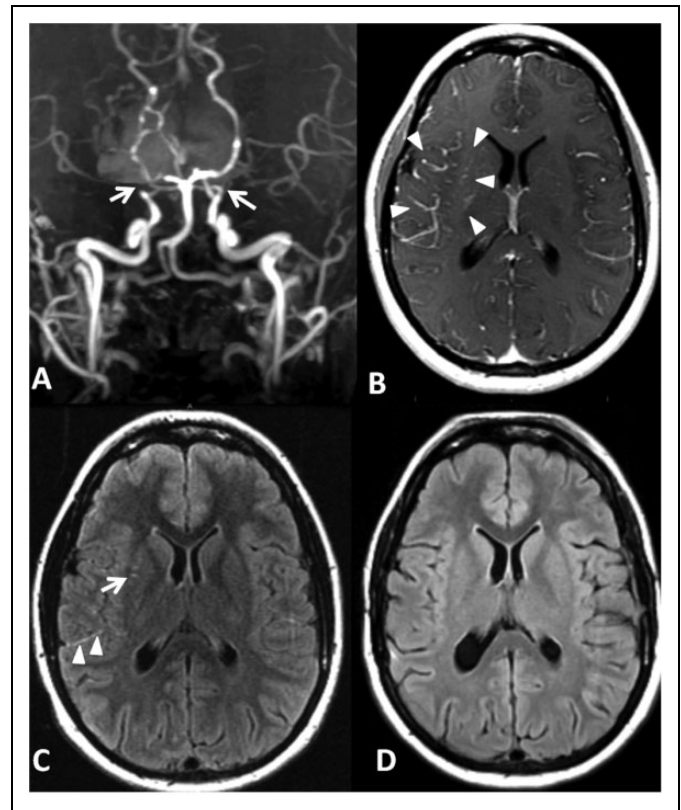


Figure 1. MRI of patient 1 demonstrating classic moyamoya findings with postoperative resolution. 3D time of flight magnetic resonance angiography demonstrates marked narrowing of the bilateral internal carotid artery (A, arrows). Contrast-enhanced and fluid-attenuated inversion recovery images show the classical “ivy sign” (B, C, arrowheads) with signal intensity alterations in the right basal ganglia (C, arrow). MRI obtained after the pial synangiosis shows resolution of the “ivy sign” and basal ganglia signal intensity alterations (D). The subtle apparent difference in ventricular size is attributable to the gantry angle being slightly different between the 2 studies.

completed 4 months after symptom onset. A contrast-enhanced MRI of the brain (Figure 1C) showed scattered areas of T2 and fluid-attenuated inversion recovery hyperintensity within the bilateral frontoparietal white matter and in the right basal ganglia. The “ivy sign” (dilation of leptomeningeal collaterals) typical of moyamoya disease was apparent on post-contrast T1-weighted images (Figure 1B). Diffusion-weighted imaging was normal. Magnetic resonance angiography of the brain (Figure 1A) showed bilateral stenosis of the carotid termini and proximal middle cerebral arteries with collateral circulation noted bilaterally and enlarged lenticulostriate vessels more apparent on the right. Conventional cerebral angiogram confirmed the magnetic resonance angiography findings. A hypercoagulable evaluation was normal. She was diagnosed with dystonia secondary to moyamoya disease. She underwent bilateral pial synangiosis 5 months after her diagnosis. Follow-up imaging revealed robust neovascularization from the indirect bypasses, along with resolution of the “ivy sign” and the signal intensity alterations in the right basal ganglia (Figure 1D). As she remained free of dystonic spells immediately after

surgery, she was weaned off oxcarbazepine 15 months after surgery. The absence of recurrence of symptoms after discontinuation of oxcarbazepine, the temporal relationship between surgery and the resolution of symptoms, and the correlation between surgery and the resolution of vascular dilation in the basal ganglia on the MRI support the hypothesis that the pial synangiosis produced long-term resolution of her dystonia. She remains symptom-free as of March 2015 on topiramate and aspirin. The topiramate is being continued for headache control.

Patient 2 presented in May 2012 as a 10-year-old left-handed female with a 4-week history of choreic movements of the left face, arm and leg. The movements occurred at rest, worsened with exertion and stress, and were leading her to fall. Her birth and family histories were unremarkable. On examination she had choreic movements of her left arm and leg that became more obvious with evocative techniques such as heel, toe and tandem walking. An MRI of the brain showed right frontal white matter T2 hyperintensities with no restricted diffusion to suggest a recent stroke. The basal ganglia were normal. A magnetic resonance angiography was not performed at this time. An echocardiogram was normal. A hypercoagulable workup was negative, as was an evaluation for demyelinating disease. Her antistreptolysin O titer was 554, and she was treated with penicillin followed by sulfadiazine prophylaxis for presumed Sydenham chorea despite not meeting Jones criteria for rheumatic fever as she met no minor diagnostic criteria (though chorea is an exception to Jones criteria).¹⁸ She had no history of preceding illness, joint pain, rash, or fever. She had no acute phase reactants, and an electrocardiogram was normal. She was begun on carbamazepine for the chorea, with complete resolution of her symptoms in the next 2 months, following which she was taken off of all medications. Eight months later, her chorea recurred. Her brain MRI showed stable nonenhancing bifrontal white matter lesions. The basal ganglia remained unremarkable. No "ivy sign" was identified. Diffusion-weighted imaging was again normal. Her magnetic resonance angiography at the time revealed bilateral, left greater than right, moyamoya disease, confirmed by formal angiography. Dilated lenticulostriate arteries were noted to be present bilaterally. Two months thereafter, she underwent bilateral pial synangiosis with complete resolution of her chorea immediately after surgery. A follow-up angiogram 1 year after surgery revealed robust neovascularization, more on the right than the left. She is asymptomatic as of January 2015.

Patient 3 presented in January 2014 as a 10-year-old male with a history of attention-deficit/hyperactivity disorder and a chief complaint of 3 weeks of abnormal movements of his face, left arm and leg. He felt "funny" just before the onset of an abnormal movement, and could suppress the movement for a few seconds. He had no pain, weakness, stiffness, or paresthesias. Because of these movements, which had progressed to be constant, he was unable to dress himself, walk, or play. He had a history of streptococcal pharyngitis 3 months prior, which was treated with an adequate antibiotic course. He had had

no episodes concerning for seizure, joint pains, rash, heart palpitations, sore throat, or fever. He had a history of a single episode of left hand and left leg numbness which lasted about 30 minutes before resolving spontaneously. He had suffered a small subarachnoid hemorrhage in December 2012 after a minor closed head injury. Family history was significant for lupus in the mother, idiopathic thrombocytopenic purpura in the maternal grandmother, moyamoya disease in a maternal great-aunt, and multiple sclerosis in the paternal grandfather. On physical examination, the patient had chorea in all 4 extremities, most severe in the left arm and leg. He had difficulty standing and walking due to the chorea. He had multiple areas of ecchymosis on his lower extremities due to recent falls. The remainder of his neurological exam was normal. An evaluation for Sydenham chorea secondary to rheumatic fever revealed an antistreptolysin O titer of 400, an erythrocyte sedimentation rate of 20 mm/hr, and a normal echocardiogram. He was begun on sulfadiazine prophylaxis despite not meeting Jones criteria¹⁸ for rheumatic fever (though chorea is an exception to the criteria). His magnetic resonance angiography of the brain showed severe distal stenosis of the internal carotid and proximal middle cerebral arteries with extensive abnormal collateralization (Figure 3C); this was confirmed angiographically (Figures 2A and 2B). Diffusion-weighted imaging was normal. Dilated leptomeningeal vessels were prominent within the basal ganglia, right greater than left (Figures 3A and 3B). An acetazolamide perfusion study showed increased perfusion with acetazolamide administration in the posterior circulation, and no increase in perfusion in the anterior circulation, evidence that the internal carotid arteries were maximally dilated in an attempt to deliver adequate blood supply to the brain. He was diagnosed with moyamoya disease, with his chorea attributed to extensive basal ganglia involvement. He was started on aspirin and carbamazepine, with significant improvement in his chorea over the next month, but not resolution. He underwent bilateral pial synangiosis surgery 1 month after diagnosis, and had no chorea immediately after surgery. A magnetic resonance angiography 6 months after surgery demonstrated near-complete resolution of the pathological leptomeningeal circulation in the basal ganglia (Figure 3D). A follow-up angiogram revealed robust bilateral neovascularization (Figures 2C and 2D). He has had no recurrence of chorea, even after being weaned off of carbamazepine, as of May 2015.

Discussion

Moyamoya disease presenting as a movement disorder is rare in the pediatric population, with chorea present in approximately 4% of new diagnoses⁸ and dystonia in an even smaller percentage. Baik and Lee's⁹ review of 38 published patients and 4 of their own patients (21 of which were pediatric) with moyamoya disease and a movement disorder identified 27 (64.3%) patients with chorea, 4 with chorea and dystonia, and 4 with isolated dystonia. Pandey et al's¹⁰ review of 446 patients (118 pediatric) with moyamoya disease identified only 4 pediatric patients with movement disorders (chorea in 2,

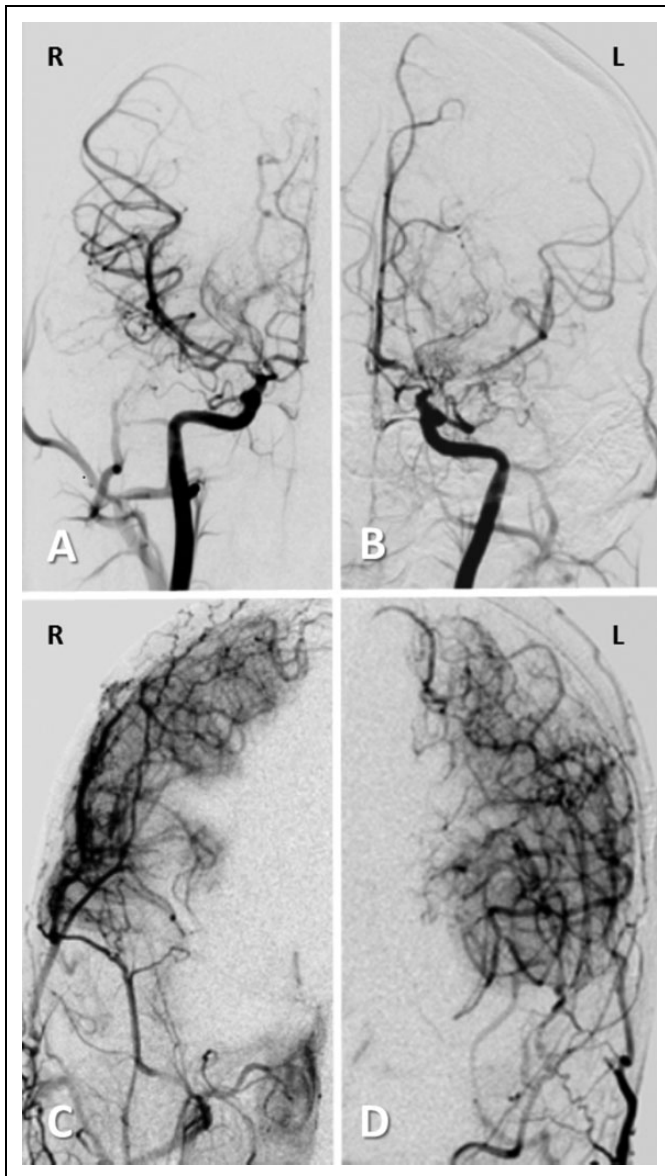


Figure 2. Angiograms on patient 3 demonstrating distal internal carotid artery stenosis and robust neovascularization postoperatively. Preoperative angiogram, selective internal carotid artery catheterization, anteroposterior view (A and B) reveals marked bilateral stenosis of the distal supraclinoid internal carotid arteries, proximal middle and anterior cerebral arteries. The left middle cerebral artery appears completely occluded. There is dilation of the lenticulostriate and anterior choroidal arteries bilaterally. Postoperative angiogram, selective external carotid catheterization, anteroposterior view (C and D) reveals significant hypertrophy of the superficial temporal arteries and extensive perfusion of the bilateral middle cerebral arterial territory.

hemiballismus in 1, and involuntary limb shaking in 1). Ahn et al¹¹ reported 9 of 316 pediatric patients whose diagnosis of chorea led to their diagnosis of moyamoya disease. Three of these patients were initially diagnosed with Sydenham's chorea. Lee et al¹⁷ described 5 pediatric patients (all with chorea, 1 with dystonia as well) with movement disorders, all of whom improved following indirect bypass.

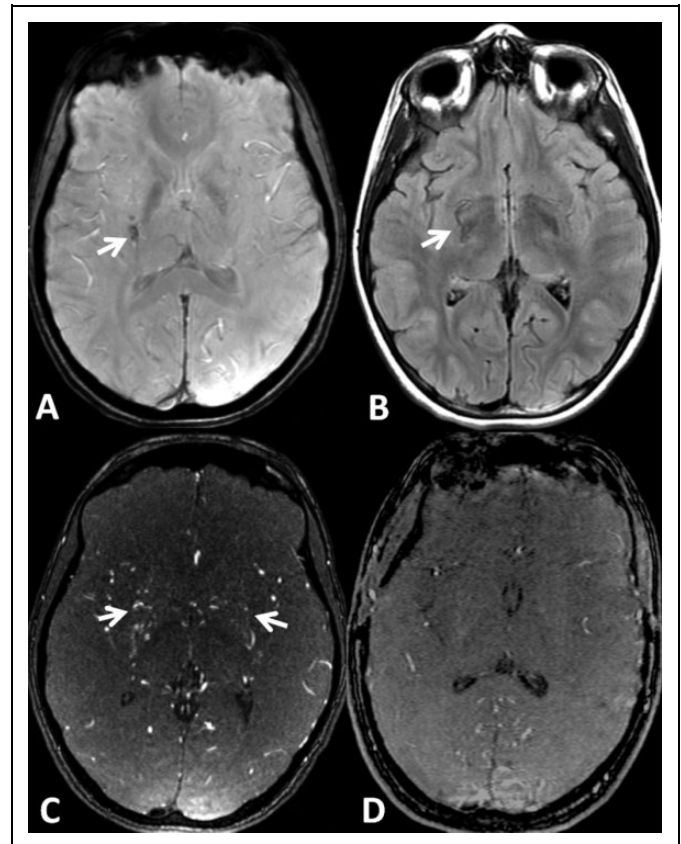


Figure 3. Dilated lenticulostriate vessels in the basal ganglia on MRI of patient 3. On susceptibility-weighted images engorgement of lenticulostriate vessels is noted in the posterior aspect of the right putamen (A, arrow). This finding is confirmed on fluid-attenuated inversion recovery images (B, arrow). 3D time of flight magnetic resonance angiography source images of the intracranial arteries obtained before pial synangiosis demonstrates prominent vessels in the bilateral basal ganglia (C, arrows) followed by complete resolution of the anomalies on the follow-up examination obtained after surgery (D).

The most frequent cause of the sudden onset of unilateral chorea or dystonia is a lacunar infarction or neoplasm in the contralateral basal ganglia (caudate, putamen, or subthalamic nucleus),¹⁹ though the most frequent cause of chorea in children is Sydenham chorea. In patients with moyamoya disease, the effect can be secondary to dilated lenticulostriate arteries compressing the basal ganglia, or due to local ischemia or infarction.¹¹ Chorea is frequently associated with lesions in the caudate nucleus or putamen, resulting in disinhibition of the external globus pallidus and hypofunction of the indirect pathway from the putamen to the internal globus pallidus. Dystonia has been correlated with isolated or combined lesions of the contralateral putamen, external globus pallidus, posterior and posterolateral thalamus, red nucleus, or subthalamic nucleus.²⁰ It correlates more strongly with increased activity of the direct pathway between the putamen and internal globus pallidus. Both pathways result in disinhibition of thalamic projections

to the premotor and motor cortex.²¹ The hypoxic-ischemic injury to the basal ganglia in moyamoya disease can be explained by selective hypoperfusion coupled with an intrinsic susceptibility of the striatum to such an insult.²⁰ The internal globus pallidus and the medial external globus pallidus are supplied by the anterior choroidal artery, whereas the caudate head and putamen (and lateral external globus pallidus) are supplied by the lenticulostriate arteries. Limited perfusion can affect either or both vascular territories.

Hypoxic injury in previously normal children and adults can cause dystonia. Bhatt et al²² reported that 10 of 12 patients with anoxic brain damage (6 children or young adults and 4 older patients) developed pure dystonia a mean of 9.7 months (range 1 week to 36 months) after anoxic events. The dystonia became generalized over a mean of 21.9 months (range 4 to 96 months). Neuroimaging generally revealed lesions in the putamen in the younger patients. Outcomes after hypoxic-ischemic injury appear to be different in infants, children, and adults.²³ In children, the delay between the insult and the movement disorder is typically longer than in adults, and the abnormal movements are far more likely to generalize.²⁴ Age-dependent changes in neuroplasticity or variability in the metabolic response of the brain to injury^{25,26} can account for these differences.

In this case series, no patient had a stroke, and all experienced resolution of their chorea or dystonia immediately following revascularization surgery. Neovascularization might not precipitate symptom resolution in the setting of infarction, though it will reduce the risk of further infarctions from upward of 90% lifetime risk to about 4% once the perioperative period has passed.⁸ The improvement of movement disorder symptoms after moyamoya surgery can be caused by normalization of cerebral autoregulation and a reduction in the degree of dilation of the lenticulostriate arteries, or by a reduction in the degree of basal ganglia ischemia. All of the authors' patients had dilated lenticulostriate arteries apparent on magnetic resonance angiography or angiography preoperatively, a finding not seen in all patients with radiographic moyamoya, and all had normalization of these arteries on follow-up imaging postoperatively. This finding, correlated with the improvement in symptoms, could be explained by either mechanical compression of the basal ganglia by the dilated lenticulostriates or by the presence of basal ganglia ischemia preoperatively, both of which would be expected to improve postoperatively. The rapid resolution of dyskinesia after indirect bypass argues against neovascularization as the key to improvement. As discussed by Lee et al,¹⁷ the pathophysiology of movement disorder in moyamoya disease remains incompletely understood.

Two of the patients (patients 2 and 3) had chorea and elevated antistreptolysin O titers, which raised the concern for Sydenham chorea and rheumatic fever. The upper limit of normal for antistreptolysin O titers is higher in children than adults and varies by geographical location; such variation is attributed to climatic or socioeconomic conditions.²⁷ The radiological findings in patient 2 and 3, followed by

complete resolution of symptoms after revascularization surgery, suggest that moyamoya disease was the etiology of their chorea. While Sydenham chorea can be the only sign of rheumatic fever in up to 20% of patients,²⁸ this must be a diagnosis of exclusion. Caution must be advised in the diagnosis of a patient with Sydenham chorea in the absence of a clear diagnosis of rheumatic fever, as occurred in both chorea patients in this series and in several patients in the Ahn series.

All 3 of the patients were treated with medication during their illnesses, a possible confounder of the conclusion. The absence of recurrence of symptoms after discontinuation of these medications, the temporal relationship between surgery and the resolution of symptoms, and the correlation between surgery and the resolution of vascular dilation in the basal ganglia on MRI and angiography support the hypothesis that pial synangiosis produced long-term resolution of their symptoms. Other authors have documented the recurrence of movement disorder symptoms when medication is stopped, evidence that medication controls the movement disorder but does not cure it.^{29,30} It is also possible that the aspirin begun to ensure graft patency, and not the surgery, produced resolution of the patients' movement disorders. However, there is no evidence that aspirin functions as a treatment for moyamoya disease, and normalization of vascular dilation in the absence of surgical treatment of moyamoya disease has not been previously reported.

Moyamoya disease must be included in the differential diagnosis of the sudden onset of dyskinesias in children and adolescents, particularly chorea and focal dystonia. In the patients with moyamoya disease and chorea, as well as in the Ahn series,¹¹ MRI/A revealed the presence of dilated lenticulostriate vessels and cerebral angiography showed the classic moyamoya disease "puff of smoke." Parenchymal infarctions can be subtle or absent on MRI, and ischemia might not be visible. The authors recommend that MRI and magnetic resonance angiography, followed by cerebral perfusion studies and possibly angiography if the initial study is abnormal, be considered in all children with the new onset of chorea or dystonia of unknown etiology to evaluate for moyamoya disease. Revascularization has demonstrated excellent results in patients with chorea in the published literature,⁹⁻¹¹ and the 2 patients with chorea and 1 with dystonia support this contention. Residual symptoms are attributable to the presence of infarction on MRI. The authors recommend that consideration be given to early surgical intervention to normalize cerebral hemodynamics and treat the movement disorder in patients with moyamoya disease.

Author Contributions

SG: study design, surgery, drafting of manuscript. LB: drafting of manuscript. KAC: review and critique of manuscript. RN: drafting of manuscript. GZ: study concept and design, interpretation of images, creation of figures, review and critique of manuscript.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

References

1. Scott RM, Smith ER. Moyamoya disease and moyamoya syndrome. *N Engl J Med*. 2009;360:1226-1237.
2. Uchino K, Johnston SC, Becker KJ, Tirschwell DL. Moyamoya disease in Washington State and California. *Neurology*. 2005;65:956-958.
3. Suzuki J, Kodama N. Moyamoya disease—a review. *Stroke J Cereb Circ*. 1983;14:104-109.
4. Kuroda S, Houkin K. Moyamoya disease: current concepts and future perspectives. *Lancet Neurol*. 2008;7:1056-1066.
5. Imaizumi T, Hayashi K, Saito K, Osawa M, Fukuyama Y. Long-term outcomes of pediatric moyamoya disease monitored to adulthood. *Pediatr Neurol*. 1998;18:321-325.
6. Imaizumi C, Imaizumi T, Osawa M, Fukuyama Y, Takeshita M. Serial intelligence test scores in pediatric moyamoya disease. *Neuropediatrics*. 1999;30:294-299.
7. Nishimoto A. [Moyamoya disease]. *Neurol Med Chir (Tokyo)*. 1979;19:221-228.
8. Scott RM, Smith JL, Robertson RL, Madsen JR, Soriano SG, Rockoff MA. Long-term outcome in children with moyamoya syndrome after cranial revascularization by pial synangiosis. *J Neurosurg*. 2004;100:142-149.
9. Baik JS, Lee MS. Movement disorders associated with moyamoya disease: a report of 4 new cases and a review of literatures. *Mov Disord*. 2010;25:1482-1486.
10. Pandey P, Bell-Stephens T, Steinberg GK. Patients with moyamoya disease presenting with movement disorder. *J Neurosurg Pediatr*. 2010;6:559-566.
11. Ahn ES, Scott RM, Robertson RL, Smith ER. Chorea in the clinical presentation of moyamoya disease: results of surgical revascularization and a proposed clinicopathological correlation. *J Neurosurg Pediatr*. 2013;11:313-319.
12. Zheng W, Wanibuchi M, Onda T, et al. A case of moyamoya disease presenting with chorea. *Childs Nerv Syst*. 2006;22(3):274-278.
13. Takanashi J, Sugita K, Honda A, Niimi H. Moyamoya syndrome in a patient with Down syndrome presenting with corea. *Pediatr Neurol*. 1993;9(5):396-398.
14. Watanabe K, Negoro T, Maehara M, Takahashi I, Nomura K, Miura K. Moyamoya disease presenting with chorea. *Pediatr Neurol*. 1990;6(1):40-42.
15. Pavlakakis SG, Schneider S, Black K, Gould RJ. Steroid-responsive chorea in moyamoya disease. *Mov Disord*. 1991;6(4):347-349.
16. Bakdash T, Cohen AR, Hempel JM, Hoagland J, Newman AJ. Moyamoya, dystonia during hyperventilation, and antiphospholipid antibodies. *Pediatr Neurol*. 2002;26(2):157-160.
17. Lee JY, Kim SK, Wang KC, et al. Involuntary movement in pediatric moyamoya disease patients: consideration of pathogenetic mechanism using neuroimaging studies. *Childs Nerv Syst*. 2014;30(5):885-890.
18. Special Writing Group of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young of the American Heart Association. Guidelines for the diagnosis of rheumatic fever. Jones Criteria, 1992 update. *JAMA*. 1992;268(15):2069-2073. Erratum: 1993;269(4):476.
19. Chuang C, Fahn S, Frucht SJ. The natural history and treatment of acquired hemidystonia: report of 33 cases and review of the literature. *J Neurol Neurosurg Psychiatry*. 2002;72:59-67.
20. Albanese A, Sorbo FD, Cornella C, et al. Dystonia rating scales: critique and recommendations. *Mov Disord*. 2013;28(7):874-883.
21. Byl NN, Merzenich MM, Jenkins WM. A primate genesis model of focal dystonia and repetitive strain injury: I. Learning-induced dedifferentiation of the representation of the hand in the primary somatosensory cortex in adult monkeys. *Neurology*. 1996;47:508-520.
22. Bhatt MH, Obeso JA, Marsden CD. Time course of postanoxic akinetic-rigid and dystonic syndromes. *Neurology*. 1993;43:314-317.
23. Burke RE, Fahn S, Gold AP. Delayed-onset dystonia in patients with “static” encephalopathy. *J Neurol Neurosurg Psychiatry*. 1980;43:789-797.
24. Saint Hilaire MH, Burke RE, Bressman SB, Brin MF, Fahn S. Delayed-onset dystonia due to perinatal or early childhood asphyxia. *Neurology*. 1991;41:216-222.
25. Rice JE, Vannucci RC, Brierley JB. The influence of immaturity on hypoxic-ischemic brain damage in the rat. *Ann Neurol*. 1981;9:131-141.
26. Vannucci RC. Mechanisms of perinatal hypoxic-ischemic brain damage. *Semin Perinatol*. 1993;17:330-337.
27. Shet A, Kaplan EL. Clinical use and interpretation of group A streptococcal antibody tests: a practical approach for the pediatrician or primary care physician. *Pediatr Infect Dis J*. 2002;21:420-430.
28. Cardoso F, Silva CE, Mota CC. Sydenham’s chorea in 50 consecutive patients with rheumatic fever. *Mov Disord*. 1997;12:701-703.
29. Harel L, Zecharia A, Straussberg R, Volovitz B, Amir J. Successful treatment of rheumatic chorea with carbamazepine. *Pediatr Neurol*. 2000;23(2):147-151.
30. Peña J, Mora E, Cardozo J, Molina O, Montiel C. Comparison of the efficacy of carbamazepine, haloperidol and valproic acid in the treatment of children with Sydenham’s chorea: clinical follow-up of 18 patients. *Arq Neuropsiquiatr*. 2002;60(2-B):374-377.