Vascular collateralization along ventriculoperitoneal shunt catheters in moyamoya disease

Technical note

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Surgically created openings such as bur holes can serve as avenues for the development of collateral blood supply to the brain in patients with moyamoya disease. When such collateralization occurs through preexisting shunt catheter sites, the potential exists for perioperative stroke if these vessels are damaged during revision of a ventricular catheter for shunt malfunction. In this paper the authors report on a series of patients with a history of ventriculoperitoneal (VP) shunts who later developed moyamoya disease and were found to have spontaneous transdural collateral vessels at ventricular catheter sites readily visualized on diagnostic angiography. A consecutive surgical series of 412 patients with moyamoya disease treated at Boston Children’s Hospital from 1990 to 2010 were reviewed to identify patients with concomitant moyamoya and a VP shunt. The clinical records and angiograms of these patients were reviewed to determine the extent of bur hole collaterals through the shunt site. Three patients were identified who had VP shunts placed for hydrocephalus and subsequently developed moyamoya disease. All 3 patients demonstrated spontaneous transdural collaterals at the ventricular catheter bur hole, as confirmed by angiography during the workup for moyamoya disease. No patients required subsequent revision of their ventricular catheters following the diagnosis of moyamoya. All patients have remained stroke free and clinically stable following pial synangiosis. Although the association of moyamoya and shunted hydrocephalus is rare, it may present a significant potential problem for the neurosurgeon treating a shunt malfunction in this patient population, because shunt bur holes may become entry sites for the ingrowth of significant cortical transdural collateral blood supply to the underlying brain. Shunt revision might therefore be associated with an increased risk of postoperative stroke or operative-site hemorrhage in this population if this vascularization is interrupted when shunt catheters are removed and replaced. A knowledge of the existence of shunt-related collaterals in patients with moyamoya may aid the surgeon in planning shunt revisions and considering, for example, a new entry point for a ventricular catheter, rather than replacing an existing one, to minimize the risk of jeopardizing existing collaterals.

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Key words • moyamoya • ventriculoperitoneal shunt • stroke • collaterals • hydrocephalus • pial synangiosis • vascular disorders

**Abbreviation used in this paper:** VP = ventriculoperitoneal.
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**Results**

Three patients were identified who had VP shunts placed for hydrocephalus at the ages of 3, 1, and 7 years, and subsequently developed moyamoya at 9, 20, and 30 years of age, respectively (Table 1). One patient (Case 2) presented with shunt malfunction before the diagnosis of moyamoya, and during surgery for shunt malfunction the proximal catheter was found to be functioning and the disconnected distal catheter was revised. All patients demonstrated spontaneous transdural collateral vessels at the ventricular catheter bur hole, as confirmed by angiography during the workup for moyamoya disease (Case 1, Fig. 1; Case 3, Fig. 2). Postoperative MRI demonstrated good collateralization at pial synangiosis sites in all patients. No patient required subsequent revision of his or her ventricular catheter following the diagnosis of moyamoya. All patients have remained stroke free and clinically stable after a mean follow-up of 5.3 years following pial synangiosis.

**Discussion**

Moyamoya disease is characterized by progressive stenosis of the intracranial internal carotid arteries and the development of collateral vessels at the apex of the carotid artery and elsewhere on the undersurface of the brain that supplies the brain distal to the stenotic vessels. Many indirect surgical revascularization procedures to treat moyamoya disease rely on the proclivity of the brain in these patients to attract the ingrowth of new collateral vessels from any adjacent vascular source.

**Table 1: Patients’ ages at the time of VP shunt placement and moyamoya diagnosis, and follow-up duration after pial synangiosis**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Cause of Hydrocephalus</th>
<th>Age at VP Shunt Placement (yrs)</th>
<th>Age at Time of Moyamoya Diagnosis (yrs)</th>
<th>Follow-Up Duration After Pial Synangiosis (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>intraventricular hemorrhage of prematurity</td>
<td>3</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>2</td>
<td>congenital hydrocephalus</td>
<td>1</td>
<td>20</td>
<td>6</td>
</tr>
<tr>
<td>3</td>
<td>craniopharyngioma</td>
<td>7</td>
<td>30</td>
<td>9</td>
</tr>
</tbody>
</table>

**Fig. 1.** Case 1. Anteroposterior (left) and lateral (right) cerebral angiograms demonstrating collateral vascularization along the ventricular catheter entry site. Arrows indicate the Rickham reservoir with collaterals and the tip of the ventricular catheter.
In this paper we report on 3 patients with a history of VP shunts who later developed moyamoya disease. These 3 patients developed good collateral vascularization along the shunt catheter tract, presumably secondary to progressing ischemia from the subsequently diagnosed moyamoya disease, and this finding is consistent with previous reports of spontaneous collateral development at bur hole sites in patients with moyamoya disease.\textsuperscript{1,2,4} There is a significant possibility that a patient with shunted hydrocephalus will require a shunt revision in the future (the rate of shunt failure after 4 years has been reported to be as high as 59\% in some series).\textsuperscript{3} In our small series, fortunately, no patients needed a shunt revision after they were diagnosed with moyamoya disease. All patients developed excellent surgical collaterals from the pial synangiosis, and it is tempting to speculate that the spontaneous collateral development from the shunt site may serve as a positive indicator of the patient’s ability to develop collaterals and thus be a good candidate for pial synangiosis.

It is important for a neurosurgeon caring for a patient with moyamoya disease and a VP shunt to understand that shunt revisions in this patient population may carry the potential risks of stroke or cerebral/subdural hemorrhage occurring during shunt catheter removal or insertion if such operative maneuvers disturb/avulse the significant preexisting brain collaterals coursing through the bur hole site. Employing strategies such as altering incisions to avoid interrupting collaterals and placing shunts at new sites while leaving old catheters alone may aid in preventing complications in patients with this rare clinical combination of moyamoya and shunted hydrocephalus. Patients with infected shunts pose a more difficult situation and careful dissection around the catheter and bur hole site to minimize disturbing the collaterals will certainly be required. Patients and their families should also be informed of the potential risk of perioperative stroke when this clinical scenario is encountered.

Patients with moyamoya disease who have VP shunts in place can develop collateral vessels along the shunt catheter tract. Physicians should be cognizant of the potential risk that these collaterals may present during shunt revision, and we recommend that neurosurgeons employ alternative shunt revision strategies to minimize these complications whenever possible.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Smith, Singla. Acquisition of data: Singla, Lin, Ho. Analysis and interpretation of data: Smith, Singla, Scott. Drafting the article: Singla, Ho. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Smith.

**References**