The Targeted Bypass Strategy for Preventing Hemorrhage in Moyamoya Disease: Technical Note

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Abstract

Although direct bypass is effective at preventing intracranial hemorrhage in moyamoya disease, the optimal strategy for achieving this purpose has rarely been addressed. The tailored targeting bypass strategy is a novel technical modification of direct bypass focused on hemorrhage prevention. The strategy is based on the promising theory of periventricular anastomosis, which explains the mechanism of hemorrhage in moyamoya disease. The strategy is defined as the use of multi-imaging modalities to predetermine in a tailored manner a target vessel at the point at which the medullary artery directly extends from the periventricular anastomosis of interest. Direct bypass with a wide craniotomy was performed on 13 hemispheres in eight patients according to this strategy. Marked shrinkage of the periventricular anastomosis of interest was observed in all but one hemisphere after surgery, and no new hemorrhages have occurred as of this writing. The present case series illustrates the technical aspects and preliminary results of the tailored targeting bypass strategy, an approach that might expand the potential of direct bypass in preventing hemorrhage.

Key words: moyamoya disease, cerebral revascularization, cerebral hemorrhage, targeting bypass

Introduction

Intracranial hemorrhage is the factor most severely affecting outcome in moyamoya disease. Hemorrhage is a serious problem in every type of this disease, as it can occur even in those initially manifesting ischemic symptom. Direct bypass is beneficial as a means of secondary prevention of hemorrhage; however, bleeding can recur even after bypass surgery. According to the Japan Adult Moyamoya Trial, about 10% of patients who had undergone bypass surgery experienced rebleeding within 5 years. To prevent hemorrhage effectively, this surgical procedure must be optimized.

“Periventricular anastomosis” is a term representing the fragile vasculature causing hemorrhage in moyamoya disease. This vasculature is characterized by the connection between the medullary artery and perforating or choroidal artery, that serves as a supply to the cortex (Fig. 1A). Periventricular vasculature can be restored to normal with successful direct bypass that eliminates pathological anastomoses via bypass flow. The improvement might be enhanced with bypass targeting the cortical area where the periventricular anastomosis is distributed.

The objective of the present technical report is to present our new tailored targeting bypass strategy, a modified direct bypass procedure for hemorrhage prevention, and its preliminary results.

Surgical Technique

Definition

“Tailored targeting bypass strategy” is defined as a technical modification of direct bypass in which surgeons select the recipient artery with reference to the distribution of the periventricular anastomosis of interest (Fig. 1B). The scalp artery is anastomosed to one of the cortical arteries (“target vessel”) at the point at which the medullary artery directly extends from the periventricular anastomosis of interest.
Patient selection

This strategy was indicated when the primary purpose of bypass surgery was hemorrhage prevention and when at least one periventricular anastomosis in the affected hemisphere apparently extended to the cortex.

Preoperative assessment

For the preoperative assessment, patients underwent magnetic resonance (MR) imaging that included whole-brain 3 Tesla time-of-flight MR angiography as well as susceptibility-weighted imaging (SWI); resting-state and acetazolamide-challenging single photon emission computed tomography (SPECT); and conventional digital subtraction angiography. Three-dimensional rotational angiography was also considered for adult patients. After imaging data was obtained, three stepwise preoperative assessments were performed:

1. Identification of periventricular anastomosis: Periventricular anastomosis was identified with sliding-thin-slab maximum-intensity-projection coronal MR angiography and conventional angiography according to the criteria set out in the Japan Adult Moyamoya Trial Group. The periventricular anastomosis was then classified into one of the three subtypes according to its origin: lenticulostriate, thalamic, or choroidal. Some hemispheres had two or three subtypes simultaneously.

2. Determination of periventricular anastomosis of interest: For patients who had suffered intracranial hemorrhage and exhibited anastomoses of multiple subtypes simultaneously, the one responsible for hemorrhage was typically the anastomosis of focus. Fusion images of SWI and the axial source image of the MR angiography were useful in determining the responsible vessel. In these images, the bleeding point was defined as the point at which the signal of the abnormally extended artery in the MRA overlapped with the hypo-intensity area in the SWI.

3. Selection of target vessel: The target vessel, a cortical artery at the point at which the periventricular anastomosis of interest directly extended, was determined through meticulous assessment of conventional and three-dimensional rotational angiography. Subsequently, MR angiography data were used to generate a brain surface image on the workstation to determine a location at which the target vessel was exposed on the surface of the brain, which represented the recipient site. This step was important because performing an anastomosis to an artery located at depth in the sulcus is difficult. The cortical veins served as good landmarks in this process. Knowledge of anatomy was also useful in determining the target vessel; a lenticulostriate anastomosis is typically distributed anterior to the central sulcus, whereas a choroidal anastomosis is typically distributed within or posterior to the central sulcus.

Operative procedure

Patients were positioned supine with the head rotated to the contralateral side. Skin incisions varied according to the number of required bypasses and the surgeons’ preference. A large craniotomy exposing the predetermined target vessel was performed to facilitate wide revascularization. After the dura was opened, the target vessel was identified on the surface of the brain with reference to a surface image generated preoperatively. For hemispheres with a periventricular anastomosis of only one subtype, we performed a single anastomosis to the target vessel, usually belonging to M4 of the middle cerebral artery (MCA), using the superficial temporal artery (STA) as a donor (STA–MCA single anastomosis). For hemispheres with multiple periventricular anastomoses of equal interest, e.g. lenticulostriate and choroidal, we considered STA–MCA double anastomosis targeting both types; alternatively, we considered single anastomosis targeting only anastomosis at higher risk of bleeding with the addition of an indirect bypass preserving the middle meningeal artery coursing above the other. For pediatric cases, encephalo-myo-synangiosis was added after anastomosis.

Postoperative assessment

Magnetic resonance imaging, SPECT, and conventional angiography were performed on all patients between 3 and 9 months (mean, 4.9 months) after
surgery to confirm the shrinkage of the periventricular anastomosis. Thereafter, patients were regularly followed with annual MR imaging including MR angiography and SWI.

Case Presentation

A 41-year-old female (Case 1 in Table 1) was diagnosed with moyamoya disease when she suffered trauma and underwent brain MRI. Although she had experienced mild transient ischemic attack in childhood, she had no apparent ischemic symptoms after reaching adulthood. Left internal carotid angiography revealed severe stenosis in the terminal portion of the internal carotid artery. It also revealed choroidal anastomosis, a connection between the anterior choroidal artery and the medullary arteries extending to the cortical branch of the MCA (Fig. 2A). An aneurysm was also observed at the site of the anastomosis. No other periventricular anastomoses were observed in the hemisphere. SPECT revealed almost normal resting-stage cerebral blood flow and reduced cerebral vascular reserve after acetazolamide challenge in the hemisphere. Although less symptomatic, the patient desired bypass surgery to reduce the hemodynamic burden on the aneurysm.

The anterior parietal artery, to which the choroidal anastomosis extended, was determined as the target vessel through angiographic assessment (Fig. 2A). The brain surface image revealed the vessel was exposed on the postcentral gyrus (Figs. 2B and 3A). A large craniotomy was performed to expose predominantly the parietal lobe (Fig. 3B). After the dura was opened, the target vessel was identified on the parietal lobe between two cortical veins (Fig. 3C). This anatomical configuration completely corresponded to that seen in the surface image generated preoperatively (Fig. 3A). The parietal branch of the STA was then anastomosed to the target vessel (Fig. 3D), and indocyanine green video angiography revealed the good patency of the bypass. The postoperative course was

| Case | Age/ Sex | Side | Symptom | Hemorrhage site | PA of interest | Direct bypass | Recipient | F/u period (months) | PA change | (Re) bleed† |
|------|----------|------|---------|----------------|----------------|---------------|-----------|-------------------|------------|-------------|
| 1    | 41/F     | L    | Asymptomatic | N/A | Choroidal | Single | Ant. parietal | 13 | Shrink | None |
| 2    | 29/F     | R    | Hemorrhage | Putamen | LSA and choroidal | Double | Prefrontal/ Central | 16 | Shrink | None |
|      |          | L    | Asymptomatic | N/A | LSA and choroidal | Double | Prefrontal/ Central | 11 | Shrink | None |
| 3    | 12/M     | R    | Hemorrhage | IVH | Choroidal | Single | Ant. parietal | 58 | Shrink | None |
| 4    | 11/M     | R    | Hemorrhage | IVH | LSA and choroidal | Single’ | Ant. parietal | 25 | Shrink | None |
|      |          | L    | Asymptomatic | N/A | LSA and choroidal | Single’ | Ant. parietal | 24 | Shrink | None |
| 5    | 65/F     | L    | Hemorrhage | Temporal | Choroidal | Single | Central | 44 | Shrink | None |
|      |          | R    | Asymptomatic | N/A | Choroidal | Single | Ant. parietal | 41 | Shrink | None |
| 6    | 9/F      | L    | Hemorrhage | IVH | Choroidal | Single | Ant. parietal | 26 | Shrink | None |
|      |          | R    | Asymptomatic | N/A | Choroidal | Single | Central | 25 | Shrink | None |
| 7    | 54/M     | R    | Hemorrhage | Temporoparietal | Choroidal | Single | Central | 13 | Persisted | None |
|      |          | L    | Asymptomatic | N/A | Choroidal | Single | Ant. parietal | 11 | Shrink | None |
| 8    | 9/F      | R    | Asymptomatic side in ischemic disease | N/A | LSA and choroidal | Single’ | Post. parietal | 11 | Shrink | None |

† Including asymptomatic bleeds detected with susceptibility weighted imaging. *Choroidal anastomosis is targeted by direct bypass, while LSA anastomosis at lower risk of bleeding is targeted by indirect bypass using the middle meningeal artery. Ant.: anterior, IVH: intraventricular hemorrhage, LSA: lenticulo striate, N/A: not available, PA: periventricular anastomosis, Post.: posterior.
uneventful. Angiography performed 9 months after surgery revealed the results: the bypass remained patent and perfused the cortex via the target vessel (Fig. 2C), and the choroidal anastomosis and aneurysm had shrunk markedly (Fig. 2D).

**Results**

The tailored targeting bypass strategy was indicated as a first-line surgical treatment for 13 hemispheres in eight cases (Table 1). In all cases, anastomosis to the intended target vessel was successful. Marked shrinkage of the periventricular anastomosis was observed in 12 of the 13 hemispheres in postoperative angiography and MR angiography. Mean follow-up period after the surgery was 24.5 months, and as of this writing no postsurgical hemorrhagic attack or increased asymptomatic bleed detected with SWI has been observed.

**Discussion**

The tailored targeting bypass strategy is a unique modification of direct bypass in that it focuses on hemorrhage prevention in moyamoya disease. The results of the Japan Adult Moyamoya Trial have revealed the effectiveness of direct bypass at preventing rebleeding. However, no study has addressed the optimal direct-bypass procedure focused on hemorrhage prevention. Although the literature suggests excellent modifications of direct bypass as well as craniotomy, most of these are focused on treating ischemic symptoms.

The seminal characteristic of tailored targeting bypass strategy is the flexibility afforded the surgeon in selecting the recipient artery to accommodate the hemorrhage mechanism. The distribution of fragile periventricular anastomoses varies by its subtype: lenticulostriate anastomoses are distributed to the
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The anterior part of the brain, whereas choroidal anastomoses are distributed to the posterior part. The surgeon may therefore select the target vessel of bypass surgery to better accommodate the subtype of periventricular anastomosis of interest. For patients with a choroidal anastomosis distributed to the parietal lobe, this strategy permits bypass even to the parietal lobe, a less likely target of conventional bypass surgery. This finding might be noteworthy because several studies have revealed that choroidal anastomosis carries an extremely high risk of bleeding. Kato et al. reported a case in which multiple aneurysms on the choroidal anastomosis had shrunk after successful direct bypass to the parietal lobe. The tailored targeting bypass strategy seems beneficial especially for those with mild hemodynamic failure, a common condition in hemorrhagic moyamoya disease. The presence of hemodynamic failure is considered the rationale for bypass surgery. The severity of hemodynamic failure, as represented by cerebral blood flow and residual cerebrovascular reserve capacity, is one factor that typically determines the extent of the bypass. Accordingly, ordinary direct bypass might be less effective for patients with mild hemodynamic failure because the distribution of bypass flow might become limited. The targeting bypass strategy might overcome this limitation because it has the potential to restore targeted periventricular vasculature directly, regardless of the extent of bypass distribution.

While the targeting bypass strategy presented here is feasible, several limitations might limit its widespread application. First, the preoperative assessment introduced in the present report often require accurate determination of the target vessel through meticulous interpretation of angiography, which can be demanding. However, the optional use of intraoperative navigation might avoid this difficulty, although the authors do not use it routinely. Second, surgeons might be concerned that the decreased number of cortical arteries, a characteristics of moyamoya disease, limits their choice of recipient artery. The theory of periventricular anastomosis, however, suggests the cortical artery is more likely to appear at the very point of distribution of periventricular anastomosis. Third, in some cases the target vessel might be invisible in the preoperative imaging, such as MR and conventional angiography, and this might impede accurate predetermination of a target vessel. In such cases, the region approximating to the distribution of the periventricular anastomosis of interest is alternatively predetermined with venous landmarks, and the cortical artery suitable for the recipient is chosen through exploration of the predetermined region during surgery.

The tailored targeting bypass strategy might expand the potential for bypass surgery to prevent hemorrhage. This strategy could also be applied as a second-line surgical treatment for patients suffering repeated hemorrhages from the medial anastomotic branch despite successful STA-MCA anastomosis. Larger and long-term follow-up studies are required to prove the efficacy of this strategy.

Conflicts of Interest Disclosure

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

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