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Management and Long-Term Outcomes of Peripheral Cerebral Aneurysms Associated with Moyamoya Disease: A Single-Center Case Series

Makoto Mizushima
Hokkaido University Graduate School of Medicine
Taku Sugiyama
takus1113@med.hokudai.ac.jp
Hokkaido University Graduate School of Medicine
Haruto Uchino
Hokkaido University Graduate School of Medicine
Masaki Ito
Hokkaido University Graduate School of Medicine
Toshiya Osanai
Hokkaido University Graduate School of Medicine
Miki Fujimura
Hokkaido University Graduate School of Medicine

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Abstract

Peripheral aneurysms arising from periventricular collateral pathways in moyamoya disease (MMD) are rare and remain challenging to manage, particularly when they persist or are detected after cerebral revascularization. We retrospectively reviewed 79 consecutive patients with MMD who experienced intracranial hemorrhage during their clinical course between 1989 and 2026 and identified 11 patients harboring 13 peripheral aneurysms. Aneurysms detected before ipsilateral revascularization were classified as pre-revascularization aneurysms (Pre-R ANs), whereas those first detected after revascularization were classified as post-revascularization aneurysms (Post-R ANs). Seven patients had Pre-R ANs and four had Post-R ANs. Among the seven patients with Pre-R ANs, six ultimately underwent combined cerebral revascularization; complete aneurysm resolution occurred in three, whereas aneurysms persisted in the remaining three. All three patients with Persistent Pre-R AN subsequently experienced rebleeding 4 months to 11 years after revascularization and underwent parent artery occlusion using N-butyl-2-cyanoacrylate (NBCA), achieving complete aneurysm obliteration without procedure-related neurological deterioration. Among the four patients with Post-R ANs, two had persistent hemodynamic insufficiency and achieved complete aneurysm resolution after additional revascularization. Of the remaining two, one underwent neuroendoscope-assisted parent artery trapping and the other showed spontaneous aneurysm resolution during observation. At final imaging follow-up, all aneurysms had resolved or been obliterated in 10 of the 11 patients. Persistent Pre-R ANs warrant careful long-term surveillance because of the potential for delayed rebleeding. For Post-R ANs, angiographic and hemodynamic assessment of revascularization adequacy may help guide treatment selection between additional revascularization and aneurysm-directed treatment.

Introduction

Moyamoya disease (MMD) is a chronic occlusive cerebrovascular disorder marked by progressive stenosis of the terminal internal carotid arteries and the compensatory formation of fragile collateral vessels at the base of the brain[7, 30]. These abnormal collateral pathways, particularly periventricular anastomoses such as choroidal and thalamic collaterals, are exposed to chronic hemodynamic stress and may occasionally give rise to peripheral aneurysms, which can result in devastating intracranial hemorrhage[9]. A systematic review of 72 patients with ruptured choroidal collateral aneurysms found that 40% of those initially managed conservatively experienced rehemorrhage, with 63% of these events occurring within 35 days; overall, 41% died or remained permanently disabled[35]. The Japan Adult Moyamoya (JAM) Trial provided randomized evidence supporting direct revascularization surgery for adult hemorrhagic MMD, with its findings suggesting a reduction in the risk of rebleeding compared with conservative management [6, 26]. However, the trial was not designed to address aneurysm-specific management and did not provide detailed information on the presence, characteristics, or clinical course of peripheral aneurysms. Consequently, the optimal management of hemorrhagic MMD complicated by peripheral aneurysms, particularly the indications and timing for direct aneurysm treatment, remains uncertain.

Several studies have suggested that cerebral revascularization alone, even in the absence of direct aneurysm obliteration procedures, may induce regression of peripheral aneurysms in MMD by alleviating hemodynamic stress on fragile collateral vessels[11, 21, 28]. However, regression of peripheral aneurysms after revascularization is not universal; some aneurysms have been reported to remain stable or persist despite successful bypass surgery[4, 28]. Furthermore, aneurysm enlargement, rerupture, or de novo formation after revascularization has also been described[20, 29, 37]. These observations suggest that cerebral revascularization may induce complex and dynamic alterations in collateral flow patterns, and that transient or localized hemodynamic stress on fragile periventricular collaterals may persist or even increase during the postoperative period[13, 37].

Parent artery occlusion using liquid embolic materials such as N-butyl-2-cyanoacrylate (NBCA) or Onyx has recently emerged as a less invasive treatment option for peripheral aneurysms that fail to regress, continue to enlarge, or cause recurrent hemorrhage after cerebral revascularization, providing an alternative to direct microsurgical intervention[19, 38, 39, 41]. However, these aneurysm-bearing arteries often supply functionally critical deep brain regions and serve as important collateral pathways in MMD. Therefore, whether parent artery occlusion can be safely performed without worsening neurological deficits must be carefully determined on a case-by-case basis, and no established criteria currently exist to guide this decision.

Although individualized treatment strategies, including additional cerebral revascularization, microsurgical aneurysm obliteration, endovascular intervention, and close radiological observation, have all been reported for peripheral aneurysms associated with MMD, these approaches are largely based on isolated case reports or small case series[1–3, 12, 29, 37]. Consequently, no evidence-based management strategy has been established, particularly for aneurysms that persist, recur, or newly develop after cerebral revascularization. Therefore, we examined the long-term clinical course and treatment outcomes of peripheral aneurysms associated with MMD to propose a practical management framework, with particular emphasis on lesions that persist or are detected after revascularization.

Materials and Methods

Ethics statement

This study was performed in accordance with the Declaration of Helsinki and its later amendments and was approved by the Institutional Review Board of Hokkaido University Hospital (approval no. 015–0086). The requirement for written informed consent was waived because only existing clinical and radiological data were used. Study information was posted on the hospital website, and patients were given the opportunity to opt out.

Study population

We retrospectively reviewed 79 consecutive patients with MMD who experienced at least one intracranial hemorrhagic event during their clinical course and were treated at Hokkaido University Hospital between January 1989 and January 2026, using our institutional MMD database.

The diagnosis of MMD was confirmed based on cerebral angiography or magnetic resonance angiography (MRA) according to the current Japanese diagnostic criteria[7, 23]. Among these patients, 11 patients (13.9%) were identified as harboring peripheral aneurysms arising from periventricular collateral pathways and were included in the present study.

Peripheral aneurysms were defined as aneurysms arising from periventricular anastomoses, including the lenticulostriate artery (LSA), thalamoperforating artery (TPA), thalamotuberal artery (TTA), anterior choroidal artery (AChA), and posterior choroidal artery (PChA)[8]. One patient included in this cohort (Case 9) was previously described in a case report of neuroendoscope-assisted aneurysm trapping for a ruptured intraventricular aneurysm associated with MMD[31]. The present study incorporates this case as part of a larger retrospective cohort analysis focused on the clinical course and management of MMD-associated peripheral aneurysms.

Data collection

Clinical and radiological records were retrospectively reviewed. The following variables were collected: sex, age at diagnosis of MMD, initial clinical presentation, age and clinical context at aneurysm detection, aneurysm side and location, number of aneurysms, age at aneurysm rupture, interval between MMD diagnosis and aneurysm rupture, Suzuki stage, history of ipsilateral cerebral revascularization before aneurysm rupture, interval between prior ipsilateral revascularization and aneurysm rupture when applicable, initial management after aneurysm rupture, postoperative development of bypasses, angiographic response to initial management, occurrence and timing of rebleeding after initial management, treatment for rebleeding, follow-up duration, final aneurysm outcome, and final functional outcome assessed using the modified Rankin Scale (mRS).

Aneurysm categorization

Given that cerebral revascularization is the mainstay of surgical treatment for MMD and may substantially alter cerebral hemodynamics, peripheral aneurysms were categorized according to the timing of detection relative to ipsilateral cerebral revascularization. Aneurysms detected before revascularization were defined as pre-revascularization aneurysms (Pre-R AN), whereas those first detected after revascularization were defined as post-revascularization aneurysms (Post-R AN). Pre-R ANs were further subclassified according to their angiographic response to revascularization into Resolved Pre-R ANs and Persistent Pre-R ANs.

As part of our routine postoperative follow-up protocol, magnetic resonance imaging (MRI) and time-of-flight magnetic resonance angiography (TOF-MRA) were generally performed in the early postoperative period and at approximately 1, 3, and 6 months. Thereafter, patients were followed with MRI and MRA at 6–12-month intervals according to their clinical condition. Postoperative development of direct and indirect bypasses was angiographically graded according to the extent of cortical revascularization, as previously described[14, 16, 34]. Revascularization was categorized as excellent, good, or poor according to the territory supplied by the surgical collaterals. The status of peripheral aneurysms was assessed for interval changes in size or morphology. Resolution was defined as complete disappearance of the aneurysm on follow-up angiographic imaging, whereas persistence was defined as failure to achieve complete disappearance. In cases in which aneurysm status could not be reliably determined by MRA, additional evaluation with digital subtraction angiography (DSA) or CT angiography (CTA) was performed.

Aneurysm response to revascularization was assessed at the lesion level, whereas clinical categorization and outcomes were assessed at the patient level. In patients with multiple Pre-R ANs, patients with at least one persistent aneurysm were classified as having Persistent Pre-R AN.

Treatment strategy

Treatment decisions were made according to the standards of care and Japanese guidelines available at the time of treatment and were individualized according to the clinical and hemodynamic status of each patient[7].

In patients with Pre-R AN, combined cerebral revascularization was generally performed as the first-line treatment without immediate aneurysm-directed intervention. Our combined revascularization procedure typically consisted of direct superficial temporal artery–middle cerebral artery (STA–MCA) bypass using one or two cortical M4 recipient arteries, together with indirect revascularization. The surgical technique and perioperative management protocol have been described in detail in our previous reports[5, 17, 22].

For Persistent Pre-R ANs and Post-R ANs, subsequent treatment was individualized on the basis of angiographic and hemodynamic findings. Cerebral angiography was used to assess bypass patency, the perfusion territory supplied by the established surgical collaterals, and the residual periventricular collateral pathway harboring the aneurysm. N-isopropyl-p-[^{123}I]iodoamphetamine (^{123}I-IMP) single-photon emission computed tomography (SPECT) was used to assess residual cerebral hemodynamic compromise. These findings were integrated to determine whether the aneurysm-bearing territory remained hemodynamically dependent on the periventricular collateral pathway.

When residual hemodynamic compromise was present and further bypass surgery was considered feasible, additional revascularization was considered[18, 33]. Aneurysm-directed treatment, primarily endovascular parent artery occlusion using NBCA, was considered when additional revascularization was technically infeasible or unlikely to provide further hemodynamic benefit, or when the aneurysm reruptured despite prior revascularization. Microsurgical or neuroendoscope-assisted trapping was considered when endovascular treatment was not feasible, whereas close observation was selected when the anticipated treatment risk was considered to outweigh the potential benefit.

Results

Patient characteristics

A total of 11 patients with MMD–associated peripheral aneurysms were identified. Patient and aneurysm characteristics are summarized in Table 1. The cohort consisted predominantly of female patients (10/11, 90.9%), with only one male patient (9.1%). The mean age at diagnosis of MMD was 42.5 ± 16.0 years. The initial clinical presentation of MMD was intracranial hemorrhage in 6 patients (54.5%), transient ischemic attack in 2 patients (18.2%), cerebral infarction in 1 patient (9.1%), and asymptomatic disease in 2 patients (18.2%). The asymptomatic cases were incidentally diagnosed during neuroimaging performed for unrelated medical conditions.

Table 1
Clinical and aneurysm characteristics of patients with MMD-associated peripheral aneurysms **Abbreviations**: F, female; M, male; mo, months; y, years; ACh choroidal artery; AN, aneurysm; CI, cerebral infarction; ICH, intracerebral hemorrhage; LPChA, lateral posterior choroidal artery; MMD, moyamoya disease; MP posterior choroidal artery; NA, not applicable or not assessable; Rt, right; Lt, left; TPA, thalamoperforating artery; TIA, transient ischemic attack.

Case No.	Sex	Age at MMD diagnosis	Initial MMD presentation	Suzuki stage	Age at AN detection	Detection context	Side / location	No. of ANs	Age at AN rupture	Interval from MMD diagnosis to AN rupture	Prior ipsilateral revascularization	Inter prior revas to AN
1	F	55	ICH	4	55	AN rupture	Rt / TPA	1	55	0	No	NA
2	F	56	ICH	3	62	AN rupture	Lt / AChA	3	62	6 y	No	NA
3	F	33	ICH	4	33	AN rupture	Lt/ AChA	1	33	0	No	NA
4	F	42	TIA	3	54	AN rupture	Lt / AChA	1	54	12 y	No	NA
5	M	37	ICH	4	37	AN rupture	Rt / LPChA	1	37	0	No	NA
6	F	54	Asymptomatic	4	54	Incidental detection	Rt / AChA	1	60	6 y	No	NA
7	F	65	ICH	3	65	Contralateral ICH unrelated to AN	Rt / AChA	1	65	0	No	NA
8	F	44	ICH	3	44	AN rupture	Rt / LPChA	1	44	9 mo	Yes	6 mo
9	F	49	Asymptomatic	3	53	AN rupture	Lt / AChA	1	53	4 y	Yes	2 y
10	F	15	CI	4	35	AN rupture	Rt / MPChA	1	35	20 y	Yes	20 y
11	F	17	TIA	4	55	AN rupture	Rt / AChA	1	55	38 y	Yes	38 y

Clinical and angiographic characteristics of peripheral aneurysms

A total of 13 peripheral aneurysms were identified in 11 patients; one patient harbored three aneurysms. With regard to aneurysm location, the anterior choroidal artery (AChA) was the most common site, accounting for 9 of the 13 aneurysms (69.2%). Three aneurysms (23.1%) arose from the posterior choroidal artery, including two from the lateral posterior choroidal artery (LPChA) and one from the medial posterior choroidal artery (MPChA). One aneurysm (7.7%) arose from the thalamoperforating artery (TPA).

Peripheral aneurysms were identified following aneurysmal hemorrhage in 9 of the 11 patients (81.8%). In one patient (9.1%), the aneurysm was incidentally detected during neuroimaging for an unrelated medical condition, whereas in another (9.1%), it was identified during evaluation of a contralateral intracerebral hemorrhage unrelated to the aneurysm. Both of these initially unruptured aneurysms subsequently ruptured during the clinical course. Seven patients (63.6%) were classified as having Pre-R AN and four (36.4%) as having Post-R AN. Among patients with Post-R AN, the interval between ipsilateral revascularization and aneurysm rupture ranged from 6 months to 38 years.

Notably, two patients with Post-R ANs (Cases 10 and 11) had ischemic presentations of MMD at 15 and 17 years of age and subsequently experienced aneurysmal hemorrhage at 35 and 55 years, respectively.

Treatment and clinical course

Management strategies and long-term outcomes are summarized in Table 2. Among the seven patients with Pre-R AN, one patient (Case 7) experienced devastating aneurysm rupture while awaiting surgery and therefore did not undergo revascularization. The remaining six patients ultimately underwent combined cerebral revascularization. Complete aneurysm resolution was achieved in three patients (Resolved Pre-R AN), whereas at least one aneurysm

persisted in the other three (Persistent Pre-R AN). In Case 2, who harbored three aneurysms, one resolved while the remaining two persisted. All three patients with Persistent Pre-R AN subsequently experienced rebleeding at 4 months, 4 years, and 11 years after revascularization, respectively. All three had well-established revascularization with no expected hemodynamic benefit from additional bypass surgery and therefore underwent parent artery occlusion using NBCA.

Table 2
Management and long-term outcomes of MMD-associated peripheral aneurysms

Case No.	Initial management after AN rupture	Bypass development after initial revascularization (direct /indirect)	AN response after initial management	Rebleeding after initial management	Interval from initial management to rebleeding	Clinical category	Treatment for rebleeding	Follow-up (y)	Final AN outcome
1	Combined revascularization	Excellent/ Excellent	Resolution	No	–	Resolved Pre-R AN	–	27.6	Resolution
2	Combined revascularization	Good/ Good	1 resolved, 2 persisted	Yes	11 y	Persistent Pre-R AN	Targeted embolization	12.6	Obliteration
3	Combined revascularization	Poor/ Excellent	Resolution	No	–	Resolved Pre-R AN	–	12.0	Resolution
4	Combined revascularization	Good/ Good	Persistence	Yes	4 mo	Persistent Pre-R AN	Targeted embolization	0.9	Obliteration
5	Combined revascularization	Good/ Excellent	Persistence	Yes	4 y	Persistent Pre-R AN	Targeted embolization	8.5	Obliteration
6	Combined revascularization	Excellent/ Poor	Resolution	No	–	Resolved Pre-R AN	–	12.8	Resolution
7	No treatment (severe deterioration)	-/-	NA	NA	NA	Pre-R AN	–	0.2	Untreated
8	Additional combined revascularization	Poor/ Poor	Resolution	No	–	Post-R AN	–	2.1	Resolution
9	Neuroendoscope-assisted AN trapping	Excellent/ Good	Obliteration	No	–	Post-R AN	–	7.6	Obliteration
10	Additional indirect revascularization	Good/ Excellent	Resolution	No	–	Post-R AN	–	5.6	Resolution
11	Observation	Good/ Excellent	Spontaneous resolution	No	–	Post-R AN	–	4.7	Resolution

Among the four patients with Post-R AN, two (Cases 8 and 10) had persistent hemodynamic insufficiency and underwent additional revascularization with occipital artery–middle cerebral artery (OA–MCA) bypass and supplementary encephalo-duro-pericranial synangiosis (EDPS), respectively, resulting in complete aneurysm resolution in both. One patient (Case 11) showed spontaneous aneurysm resolution without additional intervention. The remaining patient (Case 9), who developed recurrent hemorrhage despite well-developed direct and indirect collateral pathways, underwent neuroendoscope-assisted parent artery trapping because additional bypass surgery was considered unlikely to provide further hemodynamic benefit. All aneurysms undergoing direct aneurysm treatment were completely obliterated without procedure-related neurological deterioration.

At the final available imaging follow-up, all aneurysms had resolved or been obliterated in 10 of the 11 patients; Case 7 did not undergo definitive treatment because of severe neurological deterioration following aneurysm rupture. The median follow-up from aneurysm detection to the final clinical assessment was 7.6 years (range, 0.2–27.6 years). At the final follow-up, 7 of the 11 patients (63.6%) had a favorable functional outcome (mRS 0–2).

Representative cases

1) Case 3: Resolution of a Pre-R AN after revascularization

A 33-year-old woman presented with headache, vomiting, and impaired consciousness due to predominantly left-sided intraventricular hemorrhage (Fig. 1A). Cerebral angiography demonstrated bilateral MMD with prominent choroidal anastomoses and a 5-mm peripheral aneurysm arising from the left AChA, which was considered the source of hemorrhage (Fig. 1B, C). ^{123}I-IMP SPECT demonstrated hemodynamic compromise in the left cerebral hemisphere.

She underwent left double STA–MCA bypass combined with encephaloduro-myo-arterio-synangiosis (EDMAS). Follow-up angiography two months later demonstrated well-developed surgical collateral circulation, attenuation of the choroidal anastomosis, and complete disappearance of the AChA aneurysm

without direct aneurysm treatment (Fig. 1D, E). She subsequently underwent contralateral revascularization and remained free of recurrent hemorrhagic or ischemic events for more than 10 years, with a final mRS score of 1.

2) Case 5

3) **Persistent Pre-R AN requiring NBCA embolization**

A 37-year-old man presented with a right temporal lobe hemorrhage. Cerebral angiography demonstrated bilateral MMD and a peripheral aneurysm arising from the right LPChA, which was considered the source of hemorrhage (Fig. 2A, B). He underwent right combined cerebral revascularization at the referring hospital.

Four years later, he developed recurrent hemorrhage adjacent to the previous hematoma with intraventricular extension (Fig. 2C). Angiography demonstrated persistence of the LPChA aneurysm. The direct and indirect surgical collaterals were well developed (Fig. 2D), and ^{123}I-IMP SPECT showed no residual hemodynamic compromise in the right hemisphere outside the previously damaged temporoparietal region (Fig. 2E). Because additional revascularization was considered unlikely to provide further hemodynamic benefit, parent artery occlusion with NBCA was performed, resulting in complete aneurysm obliteration without neurological deterioration (Fig. 2F-H). No further hemorrhagic events occurred during four years of follow-up, and he remained functionally independent with a final mRS score of 2.

4) Case 8

5) **Resolution of a Post-R AN after additional revascularization**

A 44-year-old woman initially presented with intraventricular hemorrhage. Cerebral angiography demonstrated bilateral MMD with a prominent right LPChA-derived periventricular collateral pathway, but no peripheral aneurysm was identified (Fig. 3A, B). Three months later, she underwent right combined cerebral revascularization with a double STA–MCA bypass.

Six months after revascularization, she developed a right thalamic hemorrhage with intraventricular extension (Fig. 3C). Angiography demonstrated persistence of the LPChA collateral pathway and a newly detected peripheral aneurysm considered responsible for the hemorrhage (Fig. 3D). The frontal branch of the STA–MCA double bypass was occluded with limited indirect collateral development (Fig. 3E), and ^{123}I-IMP SPECT demonstrated persistent right hemispheric hemodynamic compromise (Fig. 3F). Additional revascularization with an OA–MCA bypass combined with EDMAS was therefore performed. Follow-up angiography six months later demonstrated improved intracranial perfusion and complete aneurysm resolution (Fig. 3G, H). At the last follow-up, she remained clinically stable with a final mRS score of 3.

Discussion

The present study examined the long-term clinical course and management of MMD-associated peripheral aneurysms arising from periventricular collateral pathways. Three findings are particularly noteworthy. First, cerebral revascularization resulted in complete aneurysm resolution in half of the patients with Pre-R AN who underwent surgery, supporting the established hemodynamic rationale for revascularization. Second, all three patients with Persistent Pre-R AN subsequently experienced rebleeding, occurring from 4 months to 11 years after revascularization, suggesting that persistent aneurysms should not necessarily be regarded as benign residual lesions. Third, Post-R ANs were observed under different hemodynamic conditions: some were associated with insufficient revascularization and resolved after additional bypass surgery, whereas others occurred despite well-developed surgical collateral circulation. These findings suggest that the timing of aneurysm detection relative to revascularization, together with postoperative angiographic and hemodynamic assessment, may provide a practical framework for individualized management.

Pre-R AN group: Cerebral revascularization remains the first-line treatment

Peripheral aneurysms arising from periventricular collateral pathways before ipsilateral cerebral revascularization are generally considered to result from chronic hemodynamic stress on fragile collateral vessels in MMD[21]. Importantly, previous angiographic studies have demonstrated that cerebral revascularization can induce regression of abnormal periventricular anastomoses, including choroidal and other deep collateral pathways, presumably by providing alternative cortical blood flow and reducing the dependence on these fragile collateral networks[10, 25, 36, 40]. This postoperative remodeling of the collateral circulation provides a plausible hemodynamic mechanism for the regression of peripheral aneurysms arising from these pathways. Consequently, cerebral revascularization can reduce this hemodynamic burden and may induce spontaneous aneurysm regression without direct aneurysm treatment[15, 21, 27, 28]. Previous systematic reviews have reported aneurysm resolution in approximately 75–82% of patients treated initially with cerebral revascularization, with no reported rebleeding after complete aneurysm disappearance[4, 35]. These observations support cerebral revascularization as the preferred initial treatment for ruptured Pre-R ANs when technically feasible. However, when rapidly recurrent or life-threatening hemorrhage precludes timely revascularization, urgent aneurysm-directed treatment may be considered for life-saving purposes, with careful consideration of the risk of compromising essential collateral flow.

Our findings were generally consistent with this concept. Among the six patients with Pre-R AN who ultimately underwent combined cerebral revascularization, complete aneurysm resolution was achieved in three (50%) without direct aneurysm treatment. Importantly, however, the remaining three patients had persistent aneurysms after revascularization, and all subsequently experienced rebleeding at 4 months, 4 years, and 11 years, respectively. Previous studies have reported similar residual aneurysms after revascularization, although they have generally been described simply as stable lesions. In the series reported by Ni et al., one of 13 aneurysms (7.7%) remained stable after combined revascularization[28]. Likewise, Liu et al. reported that two of five pediatric posterior choroidal artery aneurysms (40%) remained stable following indirect revascularization[24]. Furthermore, in the recent systematic review

by Feng et al., four of 27 aneurysms (14.8%) remained stable and one (3.7%) decreased in size after cerebral revascularization[4]. Importantly, however, the subsequent long-term clinical course of these persistent aneurysms has not been adequately described, and their long-term risk of rerupture remains uncertain. Although our sample size is small, the present observations suggest that persistent aneurysms after revascularization may retain a clinically relevant risk of delayed hemorrhage and therefore warrant continued vascular surveillance.

Several authors have reported primary aneurysm-directed treatment for ruptured peripheral aneurysms associated with MMD, including microsurgical trapping or clipping and endovascular parent artery occlusion, either alone or in combination with cerebral revascularization[1–3, 12, 32]. However, we believe these approaches should be adopted with caution, particularly before adequate surgical collateral circulation has been established. In the systematic review by Wiedmann et al., surgical treatment with or without revascularization was associated with relatively high complication rates, including an overall complication rate of 25% and permanent neurological deficits in 20% of patients[35]. Similar concerns have also been raised regarding endovascular treatment because the parent vessels harboring these aneurysms are often fragile periventricular collateral pathways supplying eloquent brain regions through compensatory collateral circulation. From a hemodynamic perspective, these aneurysm-bearing collateral vessels arise as a compensatory response to chronic cerebral hypoperfusion. Therefore, direct occlusion of the parent artery before sufficient surgical collateral circulation has developed may interrupt an essential collateral pathway and substantially increase the risk of ischemic complications. For this reason, we believe that direct aneurysm treatment should generally be considered only after angiographic and hemodynamic assessment confirms adequate alternative perfusion, or when further revascularization is technically infeasible or unlikely to provide additional benefit. Conversely, once angiography and cerebral blood flow studies confirm that the aneurysm-bearing collateral territory is adequately supplied by mature surgical collaterals, the hemodynamic significance of the original periventricular collateral pathway is expected to be markedly reduced. Under these circumstances, parent artery occlusion by either NBCA embolization or microsurgical trapping may be performed with a substantially lower risk of ischemic complications. Consistent with this concept, all three patients with persistent aneurysms after successful revascularization (Persistent Pre-R AN) in our series underwent parent artery occlusion using NBCA without any procedure-related neurological deterioration.

Post-R AN group: Hemodynamic evaluation–guided individualized management

Although cerebral revascularization is generally expected to reduce hemodynamic stress on periventricular collateral pathways, resulting in regression of both the collateral vessels and associated peripheral aneurysms, de novo peripheral aneurysms arising from periventricular collateral pathways have occasionally been reported after ipsilateral cerebral revascularization[20, 29]. Kim et al. reported a patient who developed a de novo distal anterior choroidal artery aneurysm 4 years after STA–MCA bypass combined with indirect revascularization[20]. The aneurysm subsequently ruptured and was treated with direct clipping. Likewise, Otawara et al. reported delayed hemorrhage from a persistent anterior choroidal artery aneurysm 10 years after bilateral STA–MCA bypass combined with indirect revascularization[29]. These reports indicate that peripheral aneurysms may newly develop or persist despite prior ipsilateral cerebral revascularization. Consistent with these previous reports, 4 of the 11 patients (36.4%) in our cohort belonged to the Post-R AN group, in whom peripheral aneurysms were first detected after ipsilateral cerebral revascularization. In our cohort, peripheral aneurysms were first detected 6 months to 38 years after ipsilateral revascularization, indicating that such lesions may become clinically apparent over a broad postoperative time course.

Our findings suggest that aneurysms first detected after ipsilateral revascularization are not a homogeneous entity but may arise under at least two distinct hemodynamic conditions: inadequate revascularization and adequate revascularization. In the former, bypass occlusion or insufficient development of surgical collaterals results in persistent cerebral hypoperfusion, leaving the aneurysm-bearing periventricular collateral pathway as an essential source of cerebral blood flow. Consequently, chronic hemodynamic stress on these fragile collateral vessels is not sufficiently relieved, predisposing to de novo aneurysm formation and subsequent rupture. This mechanism was supported by two patients in our cohort who showed residual hypoperfusion on ^{123}I-IMP SPECT despite prior revascularization. In both cases, additional targeted cerebral revascularization successfully eliminated the aneurysms, suggesting that persistent hemodynamic stress on the aneurysm-bearing periventricular collateral pathway due to inadequate bypass coverage was the principal mechanism of aneurysm formation. In contrast, the mechanism underlying de novo aneurysm formation despite adequate revascularization remains unclear. In this subgroup, both direct and indirect surgical collaterals were well developed, and ^{123}I-IMP SPECT demonstrated no apparent residual hemodynamic compromise. Nevertheless, peripheral aneurysms developed on persistent periventricular collateral pathways. Although the precise mechanism remains speculative, localized flow redistribution, persistent focal hemodynamic stress within the residual collateral network, or time-dependent alterations in postoperative cerebral blood flow may contribute to aneurysm formation. Because additional revascularization is unlikely to provide further hemodynamic benefit in these patients, direct aneurysm treatment, such as parent artery occlusion using NBCA, should be considered when appropriate.

Based on these observations, we believe that the management of de novo aneurysms after cerebral revascularization should begin with comprehensive angiographic and hemodynamic evaluation. Assessment of bypass development and collateral circulation by cerebral angiography, together with evaluation of cerebral perfusion using SPECT, may help distinguish inadequate from adequate revascularization. This distinction is clinically important because it directly influences treatment selection, favoring additional targeted revascularization in the former and aneurysm-directed treatment in the latter.

Proposed management framework

Based on the present findings and available literature, we propose the practical management framework shown in Fig. 4.

For Pre-R ANs, cerebral revascularization should generally remain the preferred initial strategy when feasible, followed by imaging surveillance to confirm aneurysm resolution. Persistent aneurysms require continued follow-up and reassessment of the established surgical collateral circulation. When adequate revascularization has been achieved and no additional hemodynamic benefit is expected from further bypass surgery, aneurysm-directed treatment may be considered.

For Post-R ANs, treatment should be guided by angiographic and hemodynamic evaluation. Persistent hypoperfusion or inadequate bypass coverage may favor additional targeted revascularization, whereas well-established surgical collateral circulation with preserved cerebral perfusion may favor aneurysm-

directed treatment. This framework should be regarded as an experience-based approach rather than a validated treatment algorithm and requires confirmation in larger cohorts.

Limitations

This study has several limitations. First, it was a retrospective, single-center case series with a small number of patients, and the observed clinical courses cannot be used to estimate the true risk of rupture or rebleeding. In particular, the finding that all three patients with Persistent Pre-R ANs subsequently experienced rebleeding should be interpreted cautiously because of the very small sample size and potential selection bias. In addition, because the cohort was restricted to patients with a history of intracranial hemorrhage, the findings may not be generalizable to peripheral aneurysms in patients with MMD who have never experienced hemorrhage. Second, the study period spanned several decades, during which surgical techniques, perioperative management, imaging protocols, and treatment strategies evolved, and procedures were performed by multiple surgeons. Treatment decisions and follow-up protocols were therefore not completely standardized. Moreover, although bypass development was graded angiographically, this grading primarily reflects the extent of cortical revascularization and does not directly quantify whether the specific territory supplied by the aneurysm-bearing collateral pathway was adequately revascularized. Third, the timing and modality of vascular follow-up varied among patients, and some aneurysms classified as Post-R AN may have been below the detection threshold on earlier imaging rather than truly developing de novo. Finally, the proposed management framework is based on a small observational cohort and should be validated in larger multicenter studies before being applied as a standardized treatment algorithm.

Conclusions

Peripheral aneurysms arising from periventricular collateral pathways in MMD exhibit heterogeneous clinical courses and should not be managed uniformly. Classification according to the timing of aneurysm detection relative to ipsilateral revascularization, together with postoperative angiographic and hemodynamic assessment, may help guide treatment selection. Cerebral revascularization should be considered the first-line treatment for ruptured peripheral aneurysms arising before ipsilateral revascularization, as it may reduce hemodynamic stress on abnormal collateral pathways and promote spontaneous aneurysm regression. Persistent Pre-R ANs warrant careful long-term surveillance because of the potential for delayed rebleeding, whereas management of Post-R ANs should be individualized according to the adequacy of cerebral revascularization.

Abbreviations

AChA
anterior choroidal artery
AN
aneurysm
CT
computed tomography
DTA
deep temporal artery
MCA
middle cerebral artery
MMA
middle meningeal artery
Pre-R
pre-revascularization
STA
superficial temporal artery.

Declarations

Ethical approval: This study was approved by the central Institutional Review Board (IRB) of Hokkaido University Hospital (approval number: 015-0086).

Clinical trial number: not applicable.

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Authors' contributions: Taku Sugiyama, Masaki Ito and Miki Fujimura contributed to the study conception and design. Data collection was performed by Makoto Mizushima, Haruto Uchino, Masaki Ito, and Toshiya Osanai. Data analysis was performed by Makoto Mizushima, Haruto Uchino, and Masaki Ito. The draft of the manuscript was written by Makoto Mizushima and critical revision was conducted by Taku Sugiyama, Haruto Uchino and Masaki Ito. All authors read and approved the final manuscript.

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References

1. Ali MJ, Bendok BR, Getch CC, Gottardi-Littell NR, Mindea S, Batjer HH (2004) Surgical management of a ruptured posterior choroidal intraventricular aneurysm associated with moyamoya disease using frameless stereotaxy: case report and review of the literature. Neurosurgery 54:1019–1024 discussion 1024. https://doi.org/10.1227/01.neu.0000115676.99378.9c
2. Chen CJ, Caruso J, Starke RM, Ding D, Buell T, Crowley RW, Liu KC (2016) Endoport-Assisted Microsurgical Treatment of a Ruptured Periventricular Aneurysm. Case Rep Neurol Med 2016:8654262. https://doi.org/10.1155/2016/8654262
3. Choulakian A, Drazin D, Alexander MJ (2010) NBCA embolization of a ruptured intraventricular distal anterior choroidal artery aneurysm in a patient with moyamoya disease. J Neurointerv Surg 2:368–370. https://doi.org/10.1136/jnis.2010.002253
4. Feng Z, Chang Y, Fu C (2024) Ruptured Peripheral Cerebral Aneurysms Associated With Moyamoya Disease: A Systematic Review. J Stroke 26:360–370. https://doi.org/10.5853/jos.2024.02061
5. Fujimura M, Tominaga T (2012) Lessons learned from moyamoya disease: outcome of direct/indirect revascularization surgery for 150 affected hemispheres. Neurol Med Chir (Tokyo) 52:327–332. https://doi.org/10.2176/nmc.52.327
6. Fujimura M, Funaki T, Houkin K, Takahashi JC, Kuroda S, Tomata Y, Tominaga T, Miyamoto S (2019) Intrinsic development of choroidal and thalamic collaterals in hemorrhagic-onset moyamoya disease: case-control study of the Japan Adult Moyamoya Trial. J Neurosurg 130:1453–1459. https://doi.org/10.3171/2017.11.Jns171990
7. Fujimura M, Tominaga T, Kuroda S, Takahashi JC, Endo H, Ogasawara K, Miyamoto S (2022) 2021 Japanese Guidelines for the Management of Moyamoya Disease: Guidelines from the Research Committee on Moyamoya Disease and Japan Stroke Society. Neurol Med Chir (Tokyo) 62:165–170. https://doi.org/10.2176/jns-nmc.2021-0382
8. Funaki T, Takahashi JC, Yoshida K, Takagi Y, Fushimi Y, Kikuchi T, Mineharu Y, Okada T, Morimoto T, Miyamoto S (2016) Periventricular anastomosis in moyamoya disease: detecting fragile collateral vessels with MR angiography. J Neurosurg 124:1766–1772. https://doi.org/10.3171/2015.6.Jns15845
9. Funaki T, Takahashi JC, Houkin K, Kuroda S, Takeuchi S, Fujimura M, Tomata Y, Miyamoto S (2018) Angiographic features of hemorrhagic moyamoya disease with high recurrence risk: a supplementary analysis of the Japan Adult Moyamoya Trial. J Neurosurg 128:777–784. https://doi.org/10.3171/2016.11.Jns161650
10. Funaki T, Miyakoshi A, Kataoka H, Takahashi JC, Takagi Y, Yoshida K, Kikuchi T, Mineharu Y, Okawa M, Yamao Y, Fushimi Y, Miyamoto S (2022) Larger Posterior Revascularization Associated with Reduction of Choroidal Anastomosis in Moyamoya Disease: A Quantitative Angiographic Analysis. AJNR Am J Neuroradiol 43:1279–1285. https://doi.org/10.3174/ajnr.A7609
11. Ge P, Ye X, Zhang Q, Liu X, Deng X, Zhao M, Wang J, Wang R, Zhang Y, Zhang D, Zhao J (2020) Clinical features, surgical treatment, and outcome of intracranial aneurysms associated with moyamoya disease. J Clin Neurosci 80:274–279. https://doi.org/10.1016/j.jocn.2020.09.006
12. Hamada J, Hashimoto N, Tsukahara T (1994) Moyamoya disease with repeated intraventricular hemorrhage due to aneurysm rupture. Report of two cases. J Neurosurg 80:328–331. https://doi.org/10.3171/jns.1994.80.2.0328
13. Hashiguchi A, Tonegawa T, Tashima K, Mizukami S, Moroki K (2024) A Case of Moyamoya Disease With Spontaneous Regression of Posterior Choroidal Artery Aneurysm After Improvement in Obesity and Administration of an Antihypertensive Drug. Cureus 16:e72458. https://doi.org/10.7759/cureus.72458
14. Ito M, Kawabori M, Sugiyama T, Tokairin K, Tatezawa R, Uchino H, Kazumata K, Houkin K, Fujimura M (2022) Impact of RNF213 founder polymorphism (p.R4810K) on the postoperative development of indirect pial synangiosis after direct/indirect combined revascularization surgery for adult Moyamoya disease. Neurosurg Rev 45:2305–2313. https://doi.org/10.1007/s10143-022-01749-9
15. Kanamori F, Takasu S, Ota S, Seki Y (2018) Prevention of the Rerupture of Collateral Artery Aneurysms on the Ventricular Wall by Early Surgical Revascularization in Moyamoya Disease: Report of Two Cases and Review of the Literature. World Neurosurg 109:393–397. https://doi.org/10.1016/j.wneu.2017.10.059
16. Kawabori M, Ito M, Kazumata K, Tokairin K, Hatanaka KC, Ishikawa S, Houkin K, Fujimura M (2023) Impact of RNF213 c.14576G > A Variant on the Development of Direct and Indirect Revascularization in Pediatric Moyamoya Disease. Cerebrovasc Dis 52:171–176. https://doi.org/10.1159/000526089
17. Kazumata K, Ito M, Tokairin K, Ito Y, Houkin K, Nakayama N, Kuroda S, Ishikawa T, Kamiyama H (2014) The frequency of postoperative stroke in moyamoya disease following combined revascularization: a single-university series and systematic review. J Neurosurg 121:432–440. https://doi.org/10.3171/2014.1.Jns13946
18. Kazumata K, Kamiyama H, Saito H, Maruichi K, Ito M, Uchino H, Nakayama N, Kuroda S, Houkin K (2017) Direct Anastomosis Using Occipital Artery for Additional Revascularization in Moyamoya Disease After Combined Superficial Temporal Artery-Middle Cerebral Artery and Indirect Bypass. Oper Neurosurg 13:213–223. https://doi.org/10.1227/neu.0000000000001346
19. Kim SH, Kwon OK, Jung CK, Kang HS, Oh CW, Han MH, Kim YS, Baik SK (2009) Endovascular treatment of ruptured aneurysms or pseudoaneurysms on the collateral vessels in patients with moyamoya disease. Neurosurgery 65:1000–1004 discussion 1004. https://doi.org/10.1227/01.Neu.0000345648.46096.Ce
20. Kim YS, Joo SP, Lee GJ, Park JY, Kim SD, Kim TS (2017) Ruptured choroidal artery aneurysms in patients with moyamoya disease: Two case series and review of the literatures. J Clin Neurosci 44:236–239. https://doi.org/10.1016/j.jocn.2017.06.055

21. Kuroda S, Houkin K, Kamiyama H, Abe H (2001) Effects of surgical revascularization on peripheral artery aneurysms in moyamoya disease: report of three cases. Neurosurgery 49:463–467 discussion 467–468. https://doi.org/10.1097/00006123-200108000-00039
22. Kuroda S, Houkin K, Ishikawa T, Nakayama N, Iwasaki Y (2010) Novel bypass surgery for moyamoya disease using pericranial flap: its impacts on cerebral hemodynamics and long-term outcome. Neurosurgery 66:1093–1101 discussion 1101. https://doi.org/10.1227/01.Neu.0000369606.00861.91
23. Kuroda S, Fujimura M, Takahashi J, Kataoka H, Ogasawara K, Iwama T, Tominaga T, Miyamoto S (2022) Diagnostic Criteria for Moyamoya Disease – 2021 Revised Version. Neurol Med Chir (Tokyo) 62:307–312. https://doi.org/10.2176/jns-nmc.2022-0072
24. Liu P, Lv XL, Liu AH, Chen C, Ge HJ, Jin HW, Feng X, Lv M, Li YX, Duan L (2016) Intracranial Aneurysms Associated with Moyamoya Disease in Children: Clinical Features and Long-Term Surgical Outcome. World Neurosurg 94:513–520. https://doi.org/10.1016/j.wneu.2016.05.039
25. Miyakoshi A, Funaki T, Takahashi JC, Takagi Y, Kikuchi T, Yoshida K, Kataoka H, Mineharu Y, Okawa M, Yamao Y, Fushimi Y, Okada T, Togashi K, Miyamoto S (2019) Restoration of periventricular vasculature after direct bypass for moyamoya disease: intra-individual comparison. Acta Neurochir (Wien) 161:947–954. https://doi.org/10.1007/s00701-019-03866-9
26. Miyamoto S, Yoshimoto T, Hashimoto N, Okada Y, Tsuji I, Tominaga T, Nakagawara J, Takahashi JC (2014) Effects of extracranial-intracranial bypass for patients with hemorrhagic moyamoya disease: results of the Japan Adult Moyamoya Trial. Stroke 45:1415–1421. https://doi.org/10.1161/strokeaha.113.004386
27. Ni W, Xu F, Xu B, Liao Y, Gu Y, Song D (2012) Disappearance of aneurysms associated with moyamoya disease after STA-MCA anastomosis with encephaloduro myosynangiosis. J Clin Neurosci 19:485–487. https://doi.org/10.1016/j.jocn.2011.05.036
28. Ni W, Jiang H, Xu B, Lei Y, Yang H, Su J, Gu Y, Mao Y (2018) Treatment of aneurysms in patients with moyamoya disease: a 10-year single-center experience. J Neurosurg 128:1813–1822. https://doi.org/10.3171/2017.3.Jns162290
29. Otawara Y, Ogasawara K, Seki K, Kibe M, Kubo Y, Ogawa A (2007) Intracerebral hemorrhage after prophylactic revascularization in a patient with adult moyamoya disease. Surg Neurol 68:335–337 discussion 337. https://doi.org/10.1016/j.surneu.2006.10.061
30. Suzuki J, Takaku A (1969) Cerebrovascular moyamoya disease. Disease showing abnormal net-like vessels in base of brain. Arch Neurol 20:288–299. https://doi.org/10.1001/archneur.1969.00480090076012
31. Tokairin K, Kazumata K, Gotoh S, Sugiyama T, Kobayashi H (2020) Neuroendoscope-Assisted Aneurysm Trapping for Ruptured Intraventricular Aneurysms in Moyamoya Disease Patients. World Neurosurg 141:278–283. https://doi.org/10.1016/j.wneu.2020.05.222
32. Tsuboki S, Mitsutake T, Kadooka K, Tanaka M (2024) Transarterial Embolization With N-Butyl-2-Cyanoacrylate for Rapidly Enlarging Distal Anterior Choroidal Artery Aneurysm Associated With Moyamoya Disease: A Case Report. Cureus 16:e65340. https://doi.org/10.7759/cureus.65340
33. Uchino H, Kashiwazaki D, Akioka N, Koh M, Kuwayama N, Houkin K, Kuroda S (2020) Strategy and effect of repeat bypass surgery for anterior/posterior circulation in refractory moyamoya disease. J Neurosurg 132:1889–1899. https://doi.org/10.3171/2019.3.Jns181979
34. Uchino H, Yamamoto S, Kashiwazaki D, Akioka N, Kuwayama N, Noguchi K, Kuroda S (2021) Using postoperative remodeling of donor arteries on MR angiography to predict the development of surgical collaterals in moyamoya disease. J Neurosurg 134:1–9. https://doi.org/10.3171/2019.8.Jns191846
35. Wiedmann MKH, Davidoff C, Lo Presti A, Ni W, Rhim JK, Simons M, Stoodley MA (2022) Treatment of ruptured aneurysms of the choroidal collateral system in moyamoya disease: a systematic review and data analysis. J Neurosurg 136:637–646. https://doi.org/10.3171/2021.1.Jns203936
36. Yamamoto S, Kashiwazaki D, Uchino H, Saito H, Hori E, Akioka N, Kuwayama N, Kuroda S (2021) Ameliorative Effects of Combined Revascularization Surgery on Abnormal Collateral Channels in Moyamoya Disease. J Stroke Cerebrovasc Dis 30:105624. https://doi.org/10.1016/j.jstrokecerebrovasdis.2021.105624
37. Yoon JT, Lee KM, Ryu J, Park JI (2024) Rapidly growing distal choroidal artery aneurysm re-rupture following revascularization for hemorrhagic Moyamoya disease: A case report. Clin Case Rep 12:e8605. https://doi.org/10.1002/ccr3.8605
38. Yu JL, Wang HL, Xu K, Li Y, Luo Q (2010) Endovascular treatment of intracranial aneurysms associated with moyamoya disease or moyamoya syndrome. Interv Neuroradiol 16:240–248. https://doi.org/10.1177/159101991001600302
39. Zhang L, Xu K, Zhang Y, Wang X, Yu J (2015) Treatment strategies for aneurysms associated with moyamoya disease. Int J Med Sci 12:234–242. https://doi.org/10.7150/ijms.10837
40. Zheng EY, Hara S, Inaji M, Tanaka Y, Nariai T, Maehara T (2023) Regression of periventricular anastomosis after indirect revascularization in pediatric patients with moyamoya disease. J Neurosurg Pediatr 32:719–728. https://doi.org/10.3171/2023.8.Peds23304
41. Zhou Z, Xu K, Yu J (2023) Parent artery occlusion for ruptured aneurysms in moyamoya vessels or on collaterals. Front Neurol 14:1085120. https://doi.org/10.3389/fneur.2023.1085120

Figures

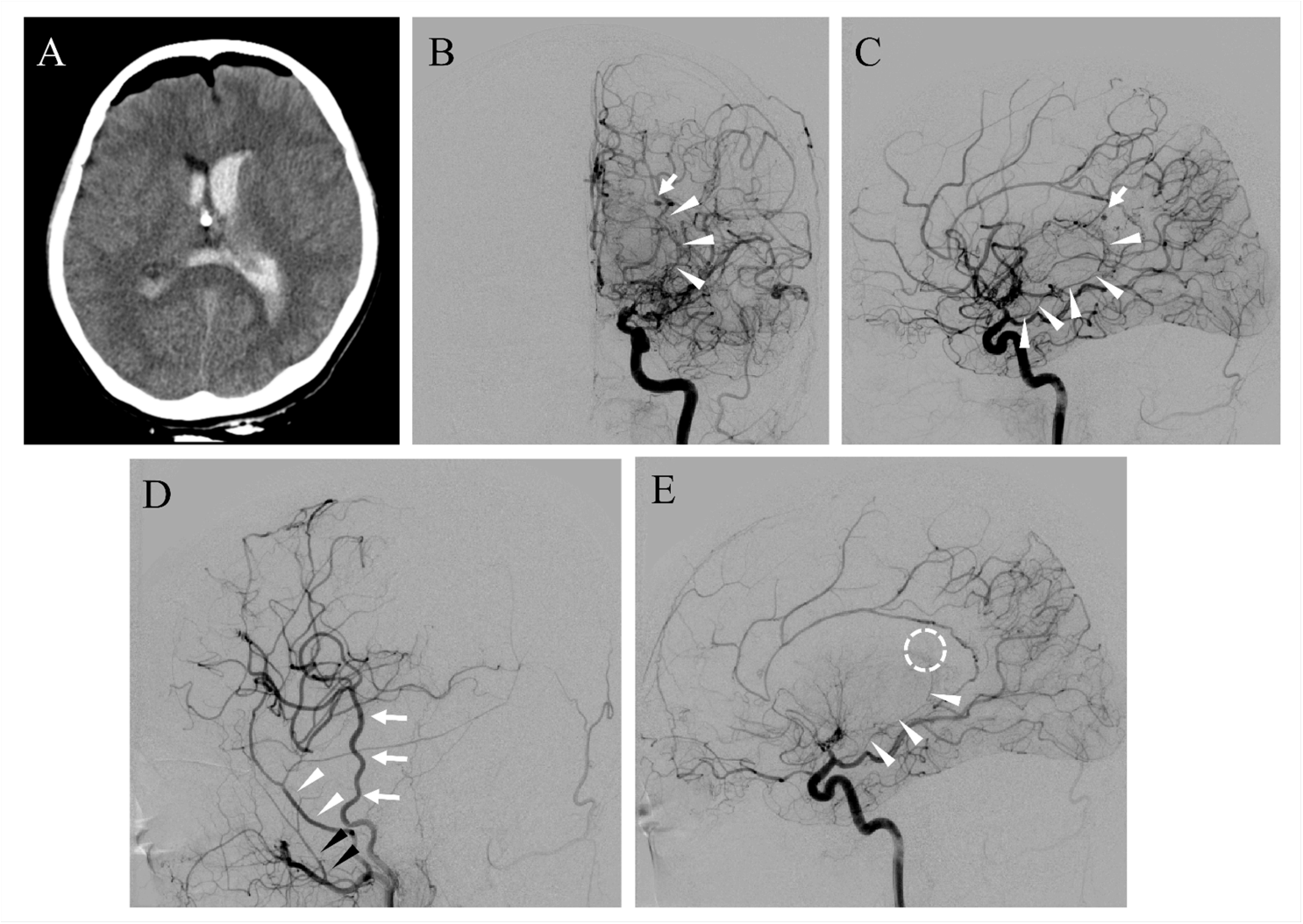


Figure 1

Case 3: Resolution of a Pre-R AN After Revascularization

(A) CT after emergency external ventricular drainage showing predominantly left-sided intraventricular hemorrhage.

(B) Frontal view of the left internal carotid angiogram demonstrating terminal internal carotid artery stenosis with a prominent choroidal anastomosis (white arrowheads) and a 5-mm peripheral aneurysm arising from the left anterior choroidal artery (white arrow).

(C) Lateral view of the left internal carotid angiogram demonstrating the choroidal anastomosis (white arrowheads) and the peripheral aneurysm (white arrow).

(D) Lateral view of the left external carotid angiogram obtained two months after surgery, showing well-developed direct STA–MCA bypass (white arrows) and indirect collateral circulation supplied by the middle meningeal artery (MMA; white arrowheads) and deep temporal artery (DTA; black arrowheads).

(E) Lateral view of the left internal carotid angiogram obtained two months after combined cerebral revascularization, showing attenuation of the choroidal anastomosis (white arrowheads) and complete resolution of the peripheral aneurysm (dotted circle).

Abbreviations:AChA, anterior choroidal artery; AN, aneurysm; CT, computed tomography; DTA, deep temporal artery; MCA, middle cerebral artery; MMA, middle meningeal artery; Pre-R, pre-revascularization; STA, superficial temporal artery.

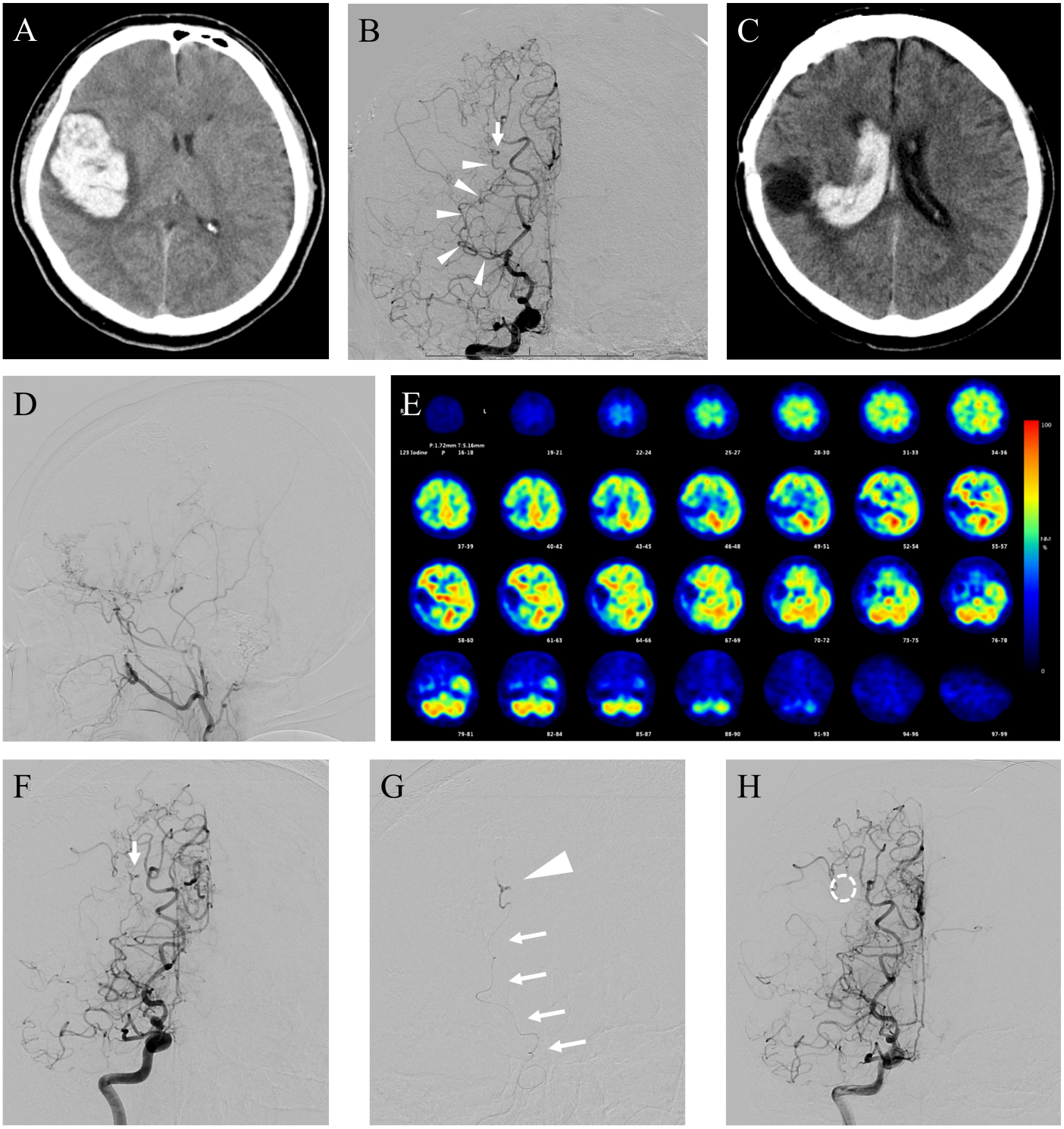


Figure 2

Abbreviations: CT, computed tomography; AN, aneurysm; LPChA, lateral posterior choroidal artery; MCA, middle cerebral artery; NBCA, N-butyl-2-cyanoacrylate; Pre-R, pre-revascularization; STA, superficial temporal artery; ^{123}I-IMP SPECT, N-isopropyl-p-[^{123}I]iodoamphetamine single-photon emission computed tomography.

Case 5: Persistent Pre-R AN Requiring NBCA Embolization

(A) CT at the initial hemorrhagic event showing a right temporal lobe hemorrhage.

(B) Right internal carotid angiogram at the initial hemorrhagic event showing a prominent LPChA-derived choroidal anastomosis (white arrowheads) and a peripheral aneurysm (white arrow).

(C) CT four years after cerebral revascularization showing recurrent hemorrhage adjacent to the previous hematoma cavity with intraventricular extension.

(D) Lateral view of the right external carotid angiogram showing a patent STA–MCA bypass and well-developed indirect collateral pathways through the middle meningeal artery and deep temporal artery.

(E) ^{123}I-IMP SPECT after the recurrent hemorrhage showing no apparent hemodynamic compromise in the right cerebral hemisphere except in the previously damaged temporoparietal region.

(F) Right internal carotid angiogram after the recurrent hemorrhage showing persistence of the LPChA aneurysm despite well-developed cerebral revascularization (white arrow).

(G) NBCA cast (white arrowhead) formed following injection through a microcatheter positioned in the right LPChA (white arrow).

(H) Right internal carotid angiogram after NBCA parent artery occlusion showing complete aneurysm obliteration (dotted circle).

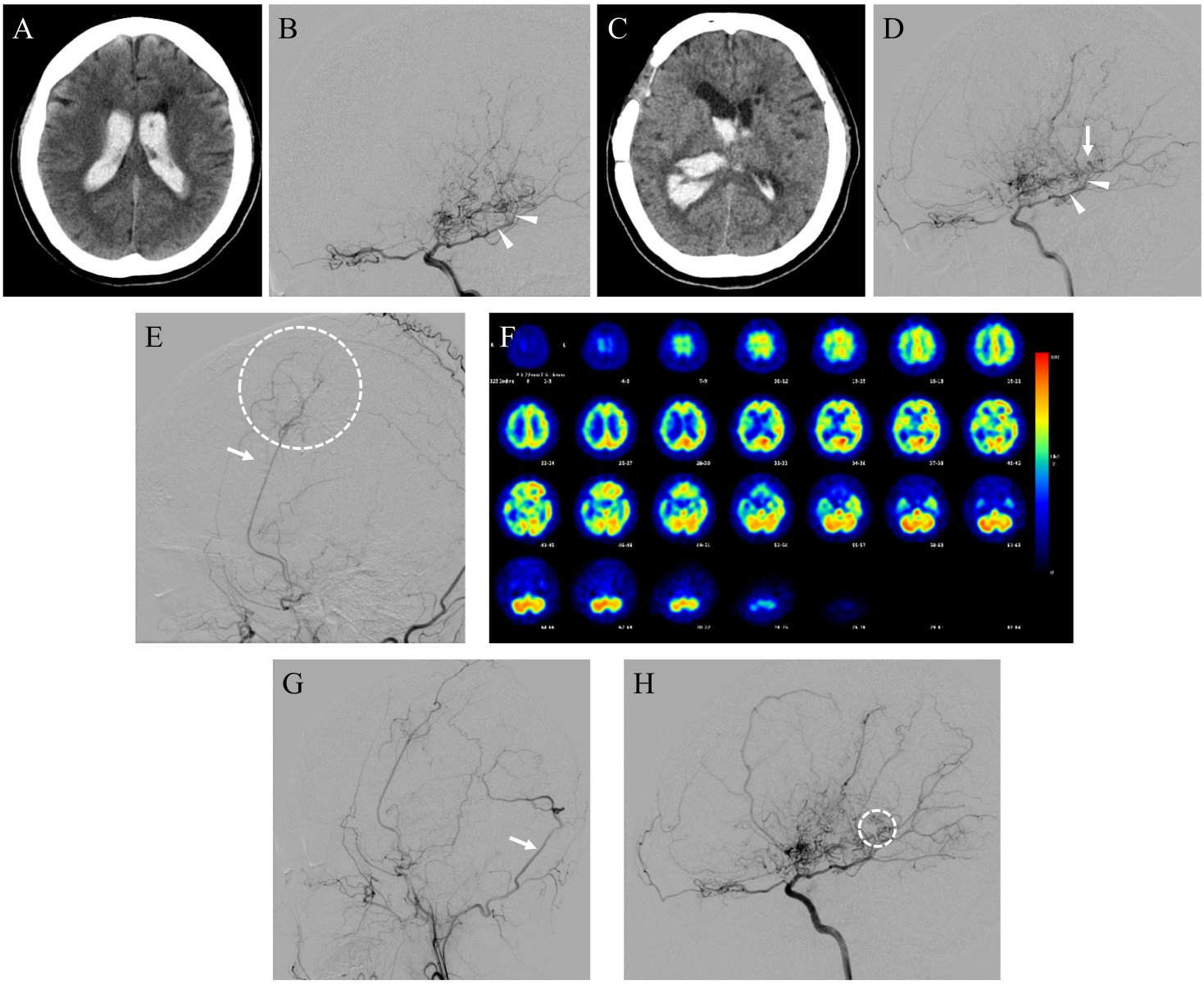


Figure 3

Case 8: Resolution of a Post-R AN After Additional Revascularization

(A) CT at the initial hemorrhagic event showing intraventricular hemorrhage.

(B) Lateral view of the right internal carotid angiogram showing a prominent LPChA-derived choroidal anastomosis (white arrowheads) without a peripheral aneurysm.

(C) CT obtained six months after right cerebral revascularization showing right thalamic hemorrhage with intraventricular extension.

(D) Lateral view of the right internal carotid angiogram after the recurrent hemorrhage showing persistence of the LPChA-derived collateral pathway (white arrowheads) and a newly detected peripheral aneurysm (white arrow).

(E) Lateral view of the right external carotid angiogram showing occlusion of the frontal branch of the STA–MCA bypass and patency of the parietal branch (white arrow), with only limited intracranial collateral perfusion through the patent parietal branch (dotted circle).

(F) ^{123}I-IMP SPECT showing hypoperfusion in the right MCA territory.

(G) Lateral view of the right external carotid angiogram showing well-developed intracranial perfusion through the OA bypass (white arrow).

(H) Lateral view of the right internal carotid angiogram obtained six months after additional OA–MCA bypass combined with EDMAS, showing complete aneurysm resolution (dotted circle).

Abbreviations: AN, aneurysm; EDMAS, encephalo-duro-myo-arterio-synangiosis; LPChA, lateral posterior choroidal artery; MCA, middle cerebral artery; OA, occipital artery; Post-R, post-revascularization; ^{123}I-IMP SPECT, N-isopropyl-p-[^{123}I]iodoamphetamine single-photon emission computed tomography; STA, superficial temporal artery.

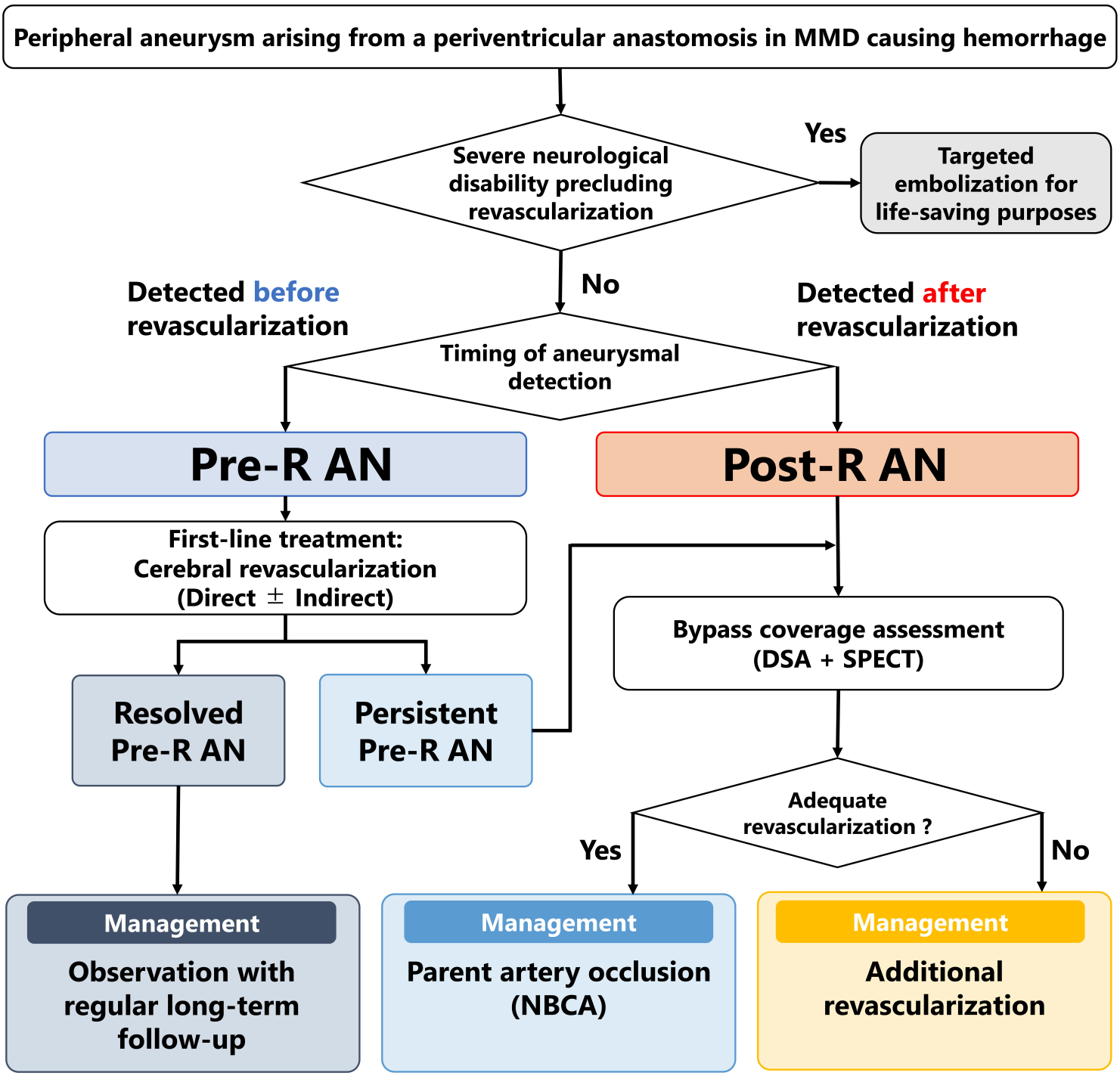


Figure 4

Proposed Management Framework for MMD-Associated Peripheral Aneurysms

Abbreviations: AN, aneurysm; MMD, moyamoya disease; NBCA, N-butyl-2-cyanoacrylate; Pre-R, pre-revascularization; Post-R, post-revascularization; DSA, digital subtraction angiography; SPECT, single-photon emission computed tomography.

Supplementary Files

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