Surgical Revascularization for Moyamoya Disease

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Clinician Reviewer: Samuel Hughes, MD, Chief of Neurosurgery

BACKGROUND

CLINICAL BACKGROUND *(excerpted from Acker 2018)*

"Moyamoya disease (MMD) is a rare cerebrovascular disease which is characterized by bilateral progressive steno-occlusion of basal cerebral arteries with emergence of coexisting abnormal net-like vessels. MMD is most frequent in Asian countries with an incidence ≤0.94/100 000, but an increase in incidence has been reported in non-Asian countries with some ethnic differences in disease characteristics. MMD shows worldwide a bimodal age distribution with a peak each in childhood and adulthood; thus, it is one of the leading causes of stroke in children and young adults. The most frequent initial symptom of MMD adults in Asians and whites is intracranial hemorrhage because of fragile blood vessels and ischemic events, respectively. Children with MMD worldwide frequently experience ischemic events."

POLICY AND CRITERIA

Coverage Guidance: Surgical revascularization for Moyamoya disease may be excluded from coverage. Check CM for exclusions or limitations.

Source	Policy
For Medicare Members	
CMS Coverage Manuals	None
National Coverage Determinations (NCD)	None
Local Coverage Determinations (LCD)	None
Local Coverage Article	None
Kaiser Permanente Medical Policy for Medicare Members	Policy 0016 applies. Due to the absence of an NCD or LCD, Kaiser Permanente has chosen to use their own Clinical Review Criteria, "Surgical Revascularization for Moyamoya Disease" for medical necessity determinations. Use the criteria below.
For Medicaid Members	
OR Medicaid	Policy 0016 does not apply
WA Medicaid	Policy 0016 does not apply
For Commercial and Self-Funded Members	
OR Commercial	Policy 0016 applies.
WA Commercial	Policy 0016 applies
Self-Funded Plans	Policy 0016 applies

Members may be eligible for revascularization surgery to treat Moyamoya disease when the following criteria are met:

1. Member has definitive Moyamoya disease as defined by ALL of the following angiographic findings:

a. Stenosis or occlusion of at least one of the following
    i. the terminal portion of the intracranial internal carotid artery;
    ii. the proximal portions of the anterior cerebral artery;
    iii. the middle cerebral artery;
b. Development of abnormal vascular networks near the occlusive or stenotic lesions in the arterial phase;
c. Bilateral cerebral lesioning; AND

2. Fulfillment of at least ONE of the following criteria:
    a. Symptoms of cerebral ischemia (e.g., ischemic stroke, transient ischemic attack, cognitive decline); OR
    b. Asymptomatic children (under 18 years of age) with:
        i. Decreased regional cerebral blood flow of less than 14%; OR
        ii. Inadequate perfusion reserve as evidenced by regional transit time greater than 8.0 seconds.

Revascularization may be direct, indirect, or a combination of both, depending upon the member's unique characteristics. Examples of indirect bypass procedures include (but are not limited to): encephaloduroarteriosynangiosis (EDAS), encephalomyosynangiosis (EDAMS), encephaloarteriosynangiosis (EAS), encephalodurogaleosynangiosis (EDGS).

RATIONALE

EVIDENCE BASIS

A comprehensive systematic review and meta-analysis (Ravindran 2019) evaluated methods of surgical revascularization among pediatric patients with Moyamoya disease. Their findings included the following:

"Of the indirect studies, a total of 488 patients were treated via encephaloduroarteriosynangliosis (EDAS), 82 via encephaloduroarteriomyosynangiosis (EDAMS), 410 via EDAS + encephalogaleosynangiosis (EGS), 216 via pial synangiosis, and 107 by dural inversion and EDAS. In the combined and direct cohort, all patients were treated with either superficial temporal artery-middle cerebral artery (STA-MCA) bypass, STA-MCA + encephalomyosynangiosis (EMS), or STA-MCA + EDAMS.

Future Stroke Incidence

The frequencies of future stroke events in patients undergoing either direct bypass alone, combined bypass, or indirect bypass alone were 1 per 190.3 patient-years, 1 per 108.9 patient-years, and 1 per 61.1 patient-years, respectively. The estimated stroke rates were 9.0% with indirect revascularization, 4.5% with direct revascularization alone, and 6.0% with combined revascularization. Stroke events most commonly occurred within the acute postoperative period, up to 7 days from surgery.

When pooling comparative studies, the overall RR of future stroke events after indirect versus combined/direct revascularization did not achieve statistical significance (RR 0.99, 95% CI 0.30–3.24, p = 0.112). On assessing the two comparative studies that included a direct bypass only, the overall RR for future stroke events after indirect versus direct bypass alone similarly did not achieve statistical significance (RR 1.84, 95% CI 0.36–9.40, p = 0.50). After pooling single-arm studies, the overall effect sizes (ESs) of the proportion of patients experiencing future stroke events were the same between combined/direct revascularization and indirect revascularization cohorts (0.04, 95% CI 0.00–0.12, and 0.04, 95% CI 0.02–0.06) and comparable with the direct bypass only cohort (0.07, 95% CI 0.03–0.16). In patients with moyamoya syndrome, the pooled postoperative stroke event rate was 6 of 102 patients (5.9%), as compared to 158 of 1864 (8.5%) in patients with idiopathic moyamoya disease.

Angiographic Outcome

The overall ESs of "excellent" angiographic outcome as designated by Matsushima grade A were 0.58 (95% CI 0.48–0.67) for indirect revascularization and 0.70 (95% CI 0.64–0.75) for combined/direct revascularization.

Complications
A total of 220 complications occurred in 1424 patients treated with indirect revascularization and 48 of 533 patients undergoing combined/direct revascularization. The most common complications in both cohorts were transient ischemic attack (TIA) and infarction within the 30-day postoperative period. Among those undergoing indirect revascularization, the 30-day ischemic infarct rate was 6.9%, relative to 2.1% in the combined/direct group. Hemorrhagic complications were similar between both groups, occurring in 1.9% of patients undergoing indirect revascularization and 0.6% of patients undergoing direct revascularization.

CODES

CPT Code	Description
61711	Anastomosis, arterial, extracranial-intracranial (e.g., middle cerebral/cortical) arteries
64999	Unlisted procedure, nervous system

ICD-10 Code and Description
I67.5 Moyamoya disease

REFERENCES

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