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Case Report

Moyamoya Disease in an 18-Month-Old Child with Down Syndrome: A Case Report, Surgical Management, and Literature Review

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Abstract

Moyamoya disease (MMD) is a chronic, progressive cerebrovascular disorder characterized by stenosis of the terminal portion of the bilateral internal carotid arteries and the formation of an abnormal collateral vascular network at the skull base. This condition was first described by Suzuki and Takaku in 1969. This case report aims to describe the surgical management and

the pathological and physiological characteristics of moyamoya disease in a child, focusing on a rare case involving an infant with Down syndrome also affected by MMD. We report an 18-month-old child who presented to the emergency department with jerky movements, brief episodes of staring or blanking, muscle weakness (lasting up to 5 minutes), rigidity, drooling, and behavioral changes. These symptoms, identified as seizures, recurred 30 minutes later. The infant also exhibited signs of right hemiparesis and a Glasgow Coma Scale (GCS) score of 9-12. Magnetic Resonance Imaging (MRI) suggested the diagnosis, which was subsequently confirmed by cerebral angiography as Moyamoya disease (MMD). The patient underwent direct extracranial-intracranial (EC-IC) revascularization. This involved a frontoparietotemporal osteoplastic craniotomy, followed by microsurgical anastomosis between the parietal branch of the left superficial temporal artery (STA) and the M4 segment of the left middle cerebral artery (MCA), supplemented with encephaloduromyoperiosynangiosis. Postoperatively, the patient developed hydrocephalus secondary to ventricular dilation. Consequently, a surgical reintervention was planned. Surgical Reintervention: During the reintervention, 350 mL of cerebrospinal fluid (CSF) was manually extracted with a syringe to reduce intracranial pressure, and an external ventricular drain (EVD) was placed intraoperatively to facilitate ongoing CSF drainage. Following the surgical reintervention, the patient was admitted to the ICU. Management included conservative treatment and antibiotics, with particular attention to patient comfort through hydration and the control of fever and hydrocephalus.

Keywords
Moyamoya disease; down syndrome; revascularization; microanastomosis; middle cerebral artery

1. Introduction

Moyamoya disease (MMD) was first described in 1957 as angiographic hypogenesis of the bilateral internal carotid arteries. However, it was formally recognized as a pathological entity by Suzuki and Takaku et al., around 1969. MMD is a chronic, progressive cerebrovascular disorder characterized by stenosis of the terminal portion of the bilateral internal carotid arteries, leading to the formation of an abnormal capillary-like vascular network at the base of the skull. This vascular pattern resembles a “puff of smoke”, which in Japanese is termed “moyamoya” [1, 2].

Moyamoya disease is the most common genetic pattern found in the R4810K polymorphism in the RNF213 ring-finger protein gene on chromosome 17q25.3; this genetic susceptibility factor is highly significant in East Asian populations [2]. Recent genetic studies have identified mutations in BRCC3/MTCP1 and GUCY1A3 as potential contributors to the pathogenesis of MMD, along with its association with certain monogenic syndromes that may underlie this angiopathy. Epidemiological data indicate that MMD has the highest prevalence in Japan, with rates ranging from 3 to 16 per 100,000 individuals annually. Research further suggests a female predominance in the affected population [3].

The Japanese Committee for Moyamoya Disease Research, established in 1974, has accumulated extensive experience in diagnosing and managing MMD, spanning over 45 years. The anatomical classification of vascular lesions dates back to 1978 [4].

The European Stroke Organization (ESO) has developed standardized guidelines for Moyamoya angiopathy, recommending direct bypass surgery in adults presenting with hemorrhagic events. Surgical intervention is typically recommended within one to two weeks following a stroke event. Additionally, some experts advocate for the use of antiplatelet therapy in non-hemorrhagic MMD to reduce the risk of embolic strokes [5].

2. Case Presentation

An 18-month-old child presented to the emergency department after several hours of jerky movements. These episodes, lasting up to 5 minutes, included brief staring spells, muscle weakness, rigidity, drooling, and behavioral changes. The seizures recurred after 30 minutes, at which point he exhibited right-sided hemiparesis and a Glasgow Coma Scale (GCS) score of 12. He was subsequently transferred to Morozoskaya Children's Hospital, where he received anticonvulsant therapy within the first hour. His medical history revealed that the seizures began after surgery for congenital heart disease. His history also included congenital Trisomy 21 (Down syndrome), multiple ventricular septal defects (perimembranous and muscular), an accessory left superior vena cava draining into the coronary sinus, tricuspid valve insufficiency, and a history of posterior cerebral artery ligation. He had previously undergone repair of a ventricular septal defect with a PTFE patch and muscular suture. Upon arrival at Morozoskaya Children's Hospital, he was administered intravenous diazepam (0.3 mL of 5 mg/solution), which successfully stopped the seizures. His Glasgow Coma Scale (GCS) score on admission to Morozoskaya Children's Hospital, in Moscow Russia, decreased to 9 to 24 hours, with persistent 2/5 right hemiparesis; magnetic resonance imaging (MRI) revealed findings suggestive of Moyamoya disease (MMD) (Figure 1). Cerebral angiography at 72 hours confirmed this diagnosis. Surgical treatment, a direct extracranial-intracranial (EC-IC) revascularization, was performed the following day. This involved a frontoparietotemporal osteoplastic craniotomy, followed by a microsurgical anastomosis between the parietal branch of the left superficial temporal artery (STA) and the M4 segment of the left middle cerebral artery (MCA) (STA-MCA), supplemented with encephaloduromyoperiosynangiosis. The direct bypass was chosen to provide immediate Revascularization. The surgery lasted 4.5 hours (Figure 2). Postoperative course: Twenty-four hours after the surgery, while in the Intensive Care Unit (ICU), the patient developed hydrocephalus secondary to ventricular dilation, necessitating a surgical reintervention. Short-term outcome: Postoperative changes: Included Enlargement of the left meningeal spaces, gas bubbles, and accumulation of CSF fluid were observed. The decision was reintervention; 350 mL of cerebrospinal fluid (CSF) was manually removed with a syringe and collected in an emesis basin or kidney tray to reduce intracranial pressure. Concurrently, an external ventricular drain (EVD) was placed intraoperatively to drain CSF and prevent further ventricular expansion and hydrocephalus recurrence. Conclusion: The patient remained in the ICU, receiving conservative treatment including antibiotics, with particular attention to comfort, hydration, fever management, and ongoing monitoring for hydrocephalus (Figure 3). Seven days later, neurological signs improved significantly, with cessation of seizures and restoration of muscle tone and reflexes. The patient was transferred to the ward with right hemiparesis improved to 3/5, and was discharged

one month later. A follow-up plan was implemented by the neurology and neurosurgery departments one month after discharge. This follow-up confirmed the positive postoperative result, with the patient remaining hemodynamically stable. See Figures 1-4.

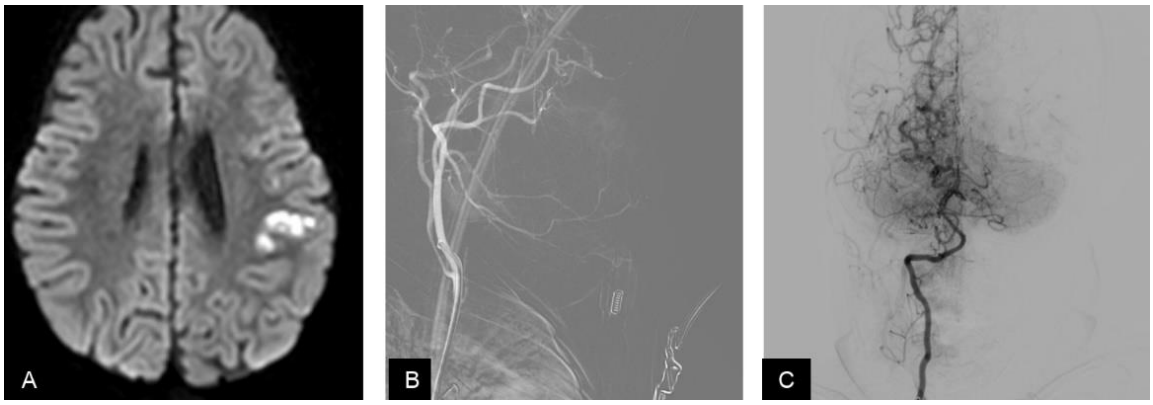

Figure 1 (A) Magnetic Resonance, an acute, large ischemic stroke is present in the left frontoparietal region. (B) Angiography showing left middle cerebral artery (MCA) territory. (C) signal is observed in the A1 segments of both anterior cerebral arteries (ACAs). A pronounced decrease in the diameter of the right internal carotid artery (ICA) accompanied by S-shaped tortuosity at the C1 level.

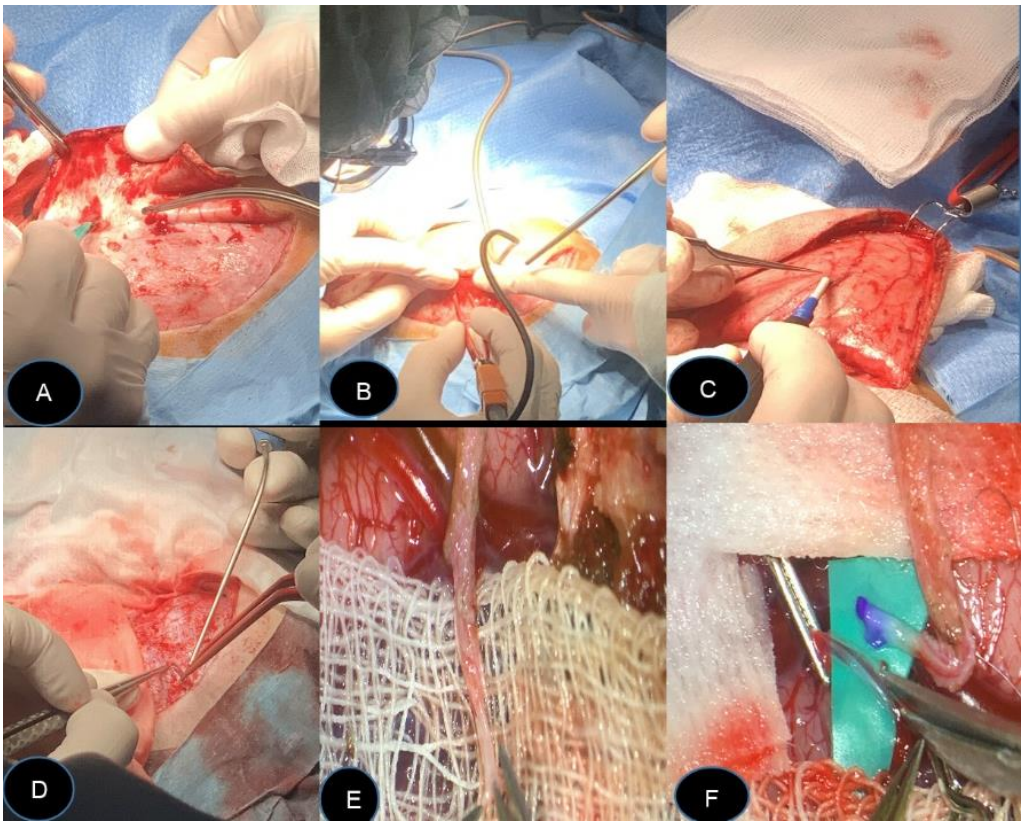

Figure 2 (A) Fronto-parieto-temporal osteoplastic craniotomy. (B) Skin-aponeurotic flap retracted to the base and fixed with hooks. (C) Coagulation of superficial vessels. (D) Microsurgical resection. (E) Microincision of the dura. (F) Extra-intracranial microanastomosis between the parietal branch of the left superficial temporal artery and the initial M4 segment of the left middle cerebral artery.

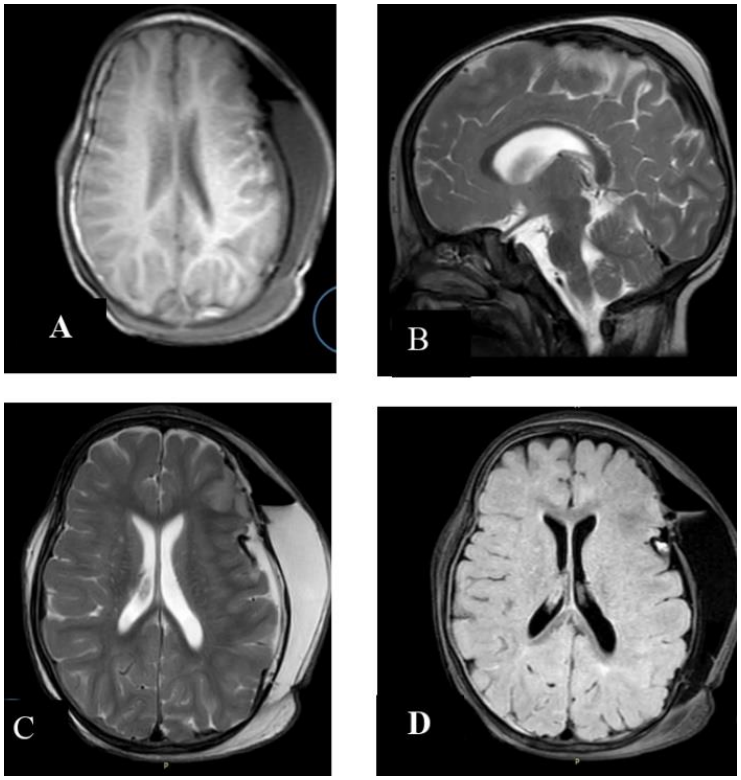

Figure 3 (A) Postoperative changes: Enlargement of the left meningeal spaces; gas bubbles are observed. (B) A sagittal image along the interhemispheric fissure reveals fluid accumulation. (C) Specifically, CSF accumulation is noted in the left parietal area. (D) In the soft tissues at the surgical site: edema, infiltration, and a gas-containing fluid collection.

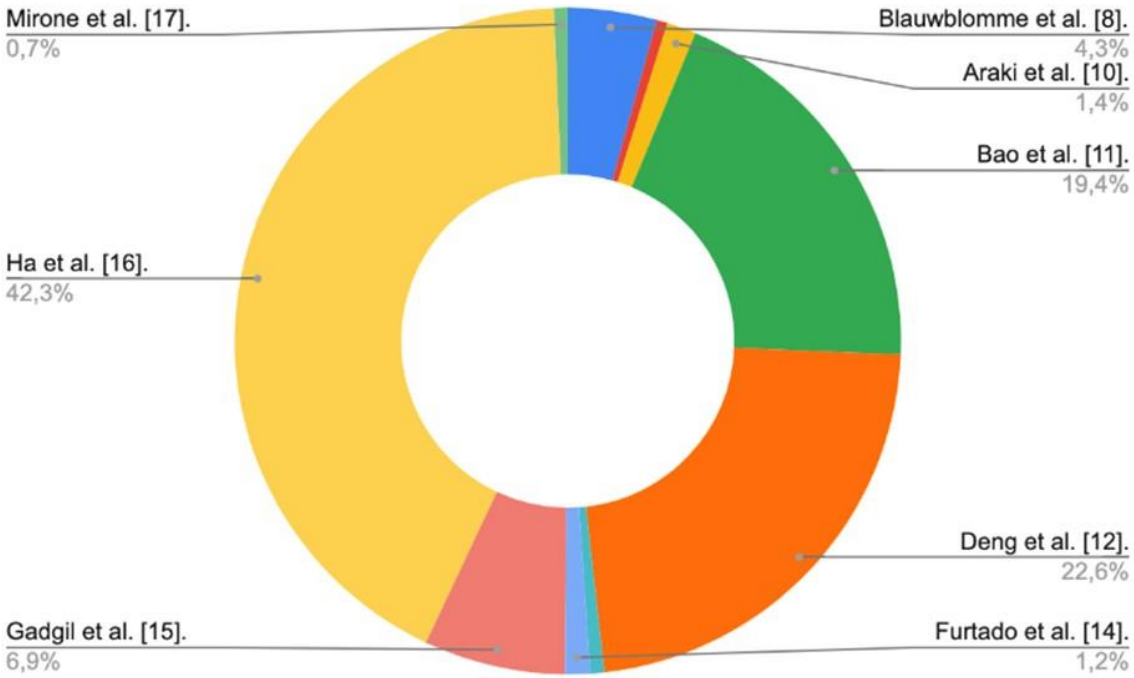

Figure 4 Recent graphical analysis, diagnostic studies, and treatments of patients with MMD using direct and indirect revascularization techniques, according to postoperative results.

3. Discussion

Moyamoya disease causes secondary ischemic stroke due to chronic stenosis of the cerebrovascular system and impaired cerebral perfusion. Revascularization surgery is the most effective treatment for establishing cerebral circulation, with direct bypass surgery particularly efficacious for optimizing perfusion of the middle cerebral artery (MCA). STA-MCA bypass is the standard direct revascularization operation to increase cerebral flow in patients with MMD [6].

This report describes an STA-MCA procedure performed on an 18-month-old child. Performing such a procedure at this young age is a complex technical decision, justified by the urgent need for immediate revascularization. Direct bypass offers advantages in critical scenarios by immediately restoring blood flow, preventing recurrent infarcts, and halting the ischemic cascade.

Neurocognitive outcomes in Moyamoya patients have been of increasing interest. Studies on Moyamoya disease have revealed that affected patients exhibit significantly lower nonverbal IQ scores, impaired fine motor function, and overall cognitive impairment on preoperative testing [7]. Despite early diagnosis, surgical revascularization remains the optimal method to reestablish cerebral perfusion and avoid subsequent ischemic injury [8] (Table 1).

Table 1 Surgical management of patients with Moyamoya disease.

Authors	Study design	Year	Patients No.	Revascularization technique	Follow up	P = value
Blauwblomme et al. [8]	Retrospective	2017	64	IB-MBH	4.2 years	<0.36
Alamri et al. [9]	Case series	2019	8	IB-EDAS	2 years	N/A
Araki et al. [10]	Retrospective	2022	21	IB-EDMS	N/A	<0.46
Bao et al. [11]	Cohort study	2015	288	IB-EDAS	2 years	N/A
Deng et al. [12]	Retrospective	2021	336	IB-EDAS, MBH	3 months recovery	<0.032
Chen et al. [13]	Cases series	2018	10	IB-Pial	63.4 months ± 36.0	N/A
Furtado et al. [14]	Cases series	2021	18	IB-EDAS	16.1 months	N/A
Gadgil et al. [15]	Cases series	2018	102	IB-EDAS	4.3 years	<0.001
Ha et al. [16]	Longitudinal and crossectional study	2019	629	IB-EDAS	12 years (range, 5-29 years)	<0.001
Mirone et al. [17]	Cases series	2019	10	IB-MBH	16 months to 7 years	N/A

IB, indirect bypass, EDAS, encephaloduroateriosynangiosis, EDMS, encephaloduromyosynangiosis, MBH multiple burr holes.

3.1 Epidemiology of Moyamoya Disease

Moyamoya disease is a rare chronic cerebrovascular disorder that can affect various ethnic groups and different countries. It is most frequently diagnosed in populations of Asian origin, followed by Black, Caucasian, and European populations. While studies analyzing patient databases in the United States show a racial distribution more proportional to the general population, the genetic component is nonetheless noteworthy. It contributes to a lower overall prevalence in Caucasian and European populations compared to East Asian regions, reflecting underlying genetic differences. In countries like Korea and Japan, family history is well reported, at 15%, while in China it is 10.7%. These populations with a family history of Moyamoya disease have a higher risk of Moyamoya disease than the general population.

Prevalence rates vary significantly by ethnicity and region. For instance, rates are 10.5 per 100,000 in Japan, 3.9 per 100,000 in China, and 16 per 100,000 in South Korea. In the United States, specifically Washington and California, the rate is 2 per 100,000. Surveillance data indicate significant increases in prevalence over time. For example, Japan saw its rate triple from 3 per 100,000 in 1995 to 10 by mid-2007, while South Korea saw more than a doubling from 2005 to 2011. This is believed to be due to improvements in non-invasive diagnostic imaging and greater access to healthcare, rather than solely to biological factors [9].

3.2 Diagnostic Criteria for Moyamoya Disease

Diagnosis of Moyamoya Disease can be established using catheter angiography and magnetic resonance angiography (MRA). The definitive conventional angiographic findings for MMD include:

1. Stenosis or occlusion in the terminal portion of the intracranial internal carotid arteries and in the proximal portions of the anterior and middle cerebral arteries.
2. The presence of abnormal vascular networks near the occlusive and stenotic lesions, observed during the arterial phase.
3. For bilateral lesions in the spontaneous occlusion stage of moyamoya disease, the 1995 MMD Diagnostic Criteria Committee determined that the criteria could be met in Japan using MRA, without requiring conventional catheter angiography. Magnetic resonance angiography (MRA) may show stenosis and occlusion of the terminal portion of the intracranial internal carotid artery (ICA) or of the proximal portions of the anterior cerebral artery (ACA) and middle cerebral artery (MCA). Additionally, when vascular networks with stenotic occlusions are present, MRA will demonstrate two or more abnormal flow patterns toward the basal ganglia in each hemisphere. These stenosis-related changes are not always evident. Although they typically originate in the terminal portion of the internal carotid artery (ICA), they may also begin in the middle cerebral artery (MCA) M1 segment. To rule out similar diseases, consider atherosclerosis, autoimmune diseases, meningitis, and brain tumors; also consider Down syndrome, neurofibromatosis type 1, head trauma, and cranial irradiation [4, 10, 14].

3.3 Moyamoya Disease with Down Syndrome

Moyamoya disease is associated with Down syndrome. In the US database, it is recorded as affecting 3.8% of hospitalized patients. Separately, 15% of patients with ischemic stroke are Caucasian or Hispanic. Patients with Down syndrome exhibit bilateral cerebral vascular deficiency,

a key characteristic of Moyamoya disease, although its pathological process is still poorly understood. Between 40% and 50% of individuals with Down syndrome have congenital cardiac defects.

Down syndrome also affects blood flow in the retinal veins that supply the optic disc. Interestingly, children with Down syndrome exhibit lower levels of systemic angiogenesis and higher levels of endostatin, which are thought to decrease the incidence of diabetic retinopathy. These angiogenic factors may also be related to other characteristics of Down syndrome, such as a low incidence of cancer [11-13]. Patients with Down syndrome may also present with thyroid dysfunction and immune dysregulation, which are linked to the development of Moyamoya disease, as this angiogenic potential may be connected to both conditions [14-16].

Moyamoya disease is associated with various genetic disorders. It can be observed in patients with neurofibromatosis type 1, where the genetic abnormality is found on chromosome 17, and in Williams syndrome, characterized by genetic mutations in the elastin gene on chromosome 7. Another example is Alagille syndrome, which involves mutations in the Jagged-1 gene. As with neurofibromatosis type 1, genes linked to Moyamoya disease may be adjacent to genes that cause these genetic pathologies [17-19]. Notably, the type IV collagen gene is located on chromosome 21, as are genes that affect arterial physiology, such as superoxide dismutase 1, interferon-Σ receptor, and cystathionine β-synthase, all of which are implicated in arterial narrowing [20-22].

In this report, our patient presents with the symptomatology of Moyamoya disease, characterized by a lack of arterial perfusion. The patient also exhibits characteristic features of Down syndrome, including a flat face and nose, a sunken chin, Mongolian hands, short neck, a single palmar crease, and upturned almond-shaped eyes. Down syndrome was diagnosed by karyotype.

3.4 Pathophysiology of Moyamoya Disease

Moyamoya disease is characterized by progressive thickening of the intimal layer, leading to stenosis and ultimately, lumen occlusion, particularly in the terminal segment of the internal carotid artery (ICA). Bilateral intimal lipid deposition contributes to this proliferative process and narrows the vessels [4]. The arteries forming the circle of Willis-including the anterior cerebral artery, middle cerebral artery, and posterior communicating artery-frequently exhibit various grades of stenosis or complete occlusion. Histologically, the arterial pathology is most often attributed to fibrocellular intimal thickening, undulation of the internal elastic lamina, and medial attenuation [15, 18, 23].

As a compensatory response, small collateral vascular plexuses develop, forming a reticular network of anastomotic branches and small perforating arteries. These reticular vascular networks, particularly within the pia mater (a thin membrane covering the brain that contains clusters of small vessels), attempt to compensate for cerebral perfusion [19, 24].

Despite these compensatory efforts, complications such as intracranial hemorrhage (initial or delayed) can occur, which may, in turn, lead to hydrocephalus. Acute hydrocephalus can be managed with external ventricular drainage, and postoperative CSF diversion can be achieved with ventriculoperitoneal shunting, which may serve as palliative treatment [20, 25].

Beyond the structural changes, genetic mutations are recognized contributors to moyamoya disease, moyamoya syndrome, and various cerebral arteriopathies. For instance, autosomal recessive moyamoya disease with achalasia can result from a mutation in the GUCY1A3 gene, which encodes a nitric oxide signaling. Heterozygous missense mutations in the ACTA2 gene are specified

as causing moyamoya disease-like symptoms by affecting the actin strands of smooth muscle. Mutations in the SAMHD1 gene are associated with an X-linked inflammatory syndrome and vasculopathy similar to moyamoya disease, often presenting with short stature and facial dysmorphia. Other identified mutations include deletions of the BRCC3 gene on the X chromosome and de novo mutations of the CBL gene, which contribute to protein degradation and severe-onset arteriopathy in pediatric moyamoya disease patients. Although the RNF213 gene was the first susceptibility gene identified, the Japanese population showed a higher prevalence of the p.R4859K mutation in Moyamoya disease. This p.R4859K variant is associated with autoimmune MMD (or "quasi-MMD") and with Moyamoya disease of arteriosclerotic origin [26].

3.5 Surgical Approach

The procedure was performed through an arcuate soft-tissue incision in the left frontoparietotemporal region. Hemostasis was achieved, and the skin-aponeurotic flap was retracted to the base and secured with surgical hooks. The parietal branch of the left STA was dissected from the skin flap over a length of 6 cm, occluded at the base with a miniclip, and irrigated with heparin solution to maintain vessel patency. A frontoparietotemporal craniotomy was performed using two trephine holes while preserving a bone bridge over the prominence of the MMA. The dura mater was incised in a petal-like fashion, carefully preserving the primary branches of the middle cerebral artery. The dural layers were placed in the subdural space beneath the margins of the bony aperture. To ensure hemostasis, the dural edges were sutured using the "Chinese roll" technique, maintaining the integrity of the principal dural branches. A microvascular anastomosis was performed between the parietal branch of the left STA (donor artery) and the M4 segment of the left middle cerebral artery (recipient artery) at the Sylvian fissure, utilizing interrupted sutures with 10/0 Prolene thread in an end-to-side configuration.

4. Conclusion

The operation in this case involved direct revascularization through an extracranial-to-intracranial bypass, connecting the superficial temporal artery to the middle cerebral artery. Microsurgical techniques were employed to increase precision and improve cerebral perfusion. Postoperative imaging showed stabilization of cerebral circulation, marked by reduced brain density loss in the frontoparietal region of the left hemisphere and improved peripheral cerebral circulation. However, a persistent area of reduced density was still observed in the left frontal lobe, with less active peripheral circulation compared to previous imaging studies.

Postoperative changes included an expansion of the membrane spaces in the operated left hemisphere to a peak of 11 mm, accompanied by gas bubbles and blood-density inclusions along the sulci and gyri. Cerebrospinal fluid accumulation was observed along the interhemispheric fissure, in the left parietal region, and in the right hemisphere. Additionally, blood accumulation was noted near the tentorium of the cerebellum on the right.

Soft-tissue changes at the operative site consisted of infiltration, edema, and a gas-filled fluid collection measuring 115 mm in length and 17 mm in thickness. The trepanation defect was adequately covered by a bone flap, which incorporated periosteum and temporalis muscle along its edge. The dural defect in the frontal region was closed using a DuraGen substitute.

Abbreviations

MMD	Moyamoya disease
ESO	The European Stroke Organization
EC-IC	Direct extracranial-intracranial revascularization
STA	Left superficial temporal artery
MCA	Middle cerebral artery
ACA	Anterior cerebral artery
EVD	External ventricular drain
CSF	Cerebrospinal fluid
ICU	Intensive care unit

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Author Contributions

Conceptualization: EBG, GC, EC, DAES; methodology: DAES, GC; software: DAES, GC; validation: ARM, GC, formal analysis: SBI, ARM; investigation: DAES; resources: EC; Data curation: SBI, AVD; writing-original draft preparation: DAES; writing-review and editing: DAES; visualization: EBG, ARM, SBI; supervision: GC, EC.

Competing Interests

The authors have declared that no competing interests exist.

AI-Assisted Technologies Statement

The authors declare that no generative AI technologies were used in the creation of this manuscript.

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