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Moyamoya disease in pediatric North African patients: a single center experience

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Keywords
Moyamoya disease, North Africa, Synangiosis, Indirect brain revascularization, Pediatric population

ABSTRACT

Introduction: Moyamoya disease (MMD) is an uncommon cerebrovascular occlusive condition characterized by progressive narrowing and obstruction of the circle of Willis. Sporadic cases have been documented in the literature from North Africa. This study presents the most comprehensive reported series of MMD cases in North Africa, emphasizing their characteristics compared with those of the largest series.

Method: We reported the clinical symptoms, imaging findings, and surgical outcomes of nine patients with Moyamoya disease using data collected from 2019 to 2022.

Results: In comparison with the Southern European population, females and pediatric patients were more frequently affected. The severity of symptom onset, neurological impairment, and vessel involvement diminished with increasing age. A review of the literature identified seven cases of Moyamoya disease (MMD) within the North African region, where authors often misdiagnosed Twig-like middle cerebral artery or Moyamoya syndrome as MMD. Statistical analysis indicates the progression of the stenosis-occlusive phenomenon within the vessel. Anterior vessels, specifically the internal carotid arteries (ICAs), are involved significantly more frequently than posterior vessels, namely the posterior cerebral arteries (PCAs; $p = 0.0009$), thereby supporting the hypothesis of anteroposterior progression. Right-to-left laterality is suggested, with a sequential progression in the following order: right ICA, left ICA, right PCA, and left PCA; however, this trend does not reach statistical significance ($p = 0.0748$).

Conclusion: Moyamoya disease is infrequent in North Africa and primarily impacts pediatric patients. Our series illustrates a distinctive anteroposterior pattern of vascular involvement and highlights the common diagnostic confusion with similar entities documented in the regional literature. These findings highlight the need for improved recognition and precise classification of MMD in this population.

INTRODUCTION

Moyamoya disease (MMD) is a rare cerebrovascular condition characterized by progressive narrowing and/or occlusion of major

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intracranial arteries. Symptoms vary with age: children often experience ischemia-related neurological episodes, while adults may present with either ischemic or hemorrhagic events.

Cerebral angiography remains the gold standard for diagnosing MMD, revealing hazy, smoke-like vessels at the skull base. For young children and toddlers, non-invasive imaging such as CTA and magnetic resonance angiography are preferred.

Although the cause is still unknown, research indicates a link to chromosome 17, which carries the MMD susceptibility gene, especially among East Asian populations. A genome-wide and locus-specific study by Kamada et al. identified RNF213 as the first gene associated with increased risk for MMD.

To prevent recurrent ischemic or hemorrhagic strokes, surgical revascularization is recommended. A combination of direct and indirect techniques is often used. This report presents a case series of nine children with MMD who underwent surgery, highlighting radiological and clinical findings, and discusses management strategies through illustrative cases.

Material and Methods

As a national moyamoya reference center, we reported 27 patients of all ages treated between January 2019 and March 2022, including 9 children with MMD and 5 with moyamoya syndrome. This study specifically focused on patients under 18 years old who were diagnosed with moyamoya disease and had no underlying conditions.

Preoperative Evaluation

Clinical and neuroradiological examinations, including preoperative magnetic resonance imaging (MRI), were used to evaluate stroke. Unilateral (right or left) versus bilateral stroke, and the number of vascular axes involved, were also differentiated.

Perfusion and diffusion sequences were studied preoperatively. However, an acetazolamide challenge test was not available.

Cerebral angiography was performed at the patient's discretion and usually in private practice if the patient could afford it.

The external carotid artery was assessed with particular attention paid to the superficial temporal, occipital, and middle meningeal arteries.

To assess global neurological impairment following stroke in children, the pediatric modified Rankin Scale (pmRS) of Bigi et al. [5] was used: (1) no symptoms; (2) no significant disabilities despite symptoms; (3) slight disability; (4) moderate disability; (5) moderately severe disability; (6) severe disability; (7) death.

Decision-Making

Following diagnosis, the primary objective of treatment in symptomatic moyamoya patients is to prevent further neurological decline resulting from ischemia or hemorrhage. Since surgical management has become the standard approach in pediatric cases, decision-making primarily centers on the timing of intervention, the selection of the appropriate technique, and the surgical side. When deemed appropriate, procedures were conducted à froid, indicating they were performed outside the acute phase of ischemic injury.

All operated patients underwent encephalo-duro-myo-synangiosis (EDMS), with some receiving additional encephalo-duro-arterio-myo-synangiosis (EDAMS) or STA-MCA bypass.

The intraoperative decision was contingent upon the disparity between the cortex and the skull, a difference that may be exacerbated by chronic or extensive infarction. Patients presenting in an unconscious state with substantial infarction were not deemed suitable candidates for surgery.

The algorithm emphasizes a structured, age- and anatomy-based approach: children over 8 years of age undergo EDMS combined with STA-MCA bypass; those aged 5-8 years receive EDAMS when the gap is less than 2 cm, and EDMS when it exceeds 2 cm; and children under 5 years are treated with EDMS. This framework provides a pragmatic guide to selecting the most appropriate revascularization strategy for moyamoya-related stroke.

Perioperative Considerations

Perioperative anesthetic management in children with moyamoya disease focused on maintaining stable cerebral perfusion and minimizing the risk of ischemic events. Cilostazol was discontinued three days before surgery to reduce intraoperative bleeding risk while preserving adequate platelet function. During patient preparation, antalgics and anxiolytics were administered to prevent crying or agitation, both of which can precipitate cerebral hypoperfusion and trigger stroke. Throughout the procedure, mean arterial pressure was carefully

maintained within 10-15% of the child's baseline, with an intentional increase of approximately 10 mmHg above baseline during temporary vessel clamping to support collateral flow. Strict normothermia and normocarbia were ensured at all times to avoid fluctuations in cerebral blood flow and metabolic demand.

Surgical Technique: EDMS

The patient was placed supine with a neutral head position on an unstabilized headrest to facilitate repositioning during the procedure. The frontal and temporal branches of the superficial temporal artery (STA) were identified and carefully marked. Standard preparation and draping were performed diligently. A skin incision was made within the superficial dermis above the marked artery under microscopic magnification, with care taken to avoid arterial injury. The temporal artery was carefully dissected to a length of 10 centimeters. Collateral branches were ligated and divided as necessary. Bipolar cauterization was reserved for uncontrollable bleeding.

The incision was extended along the upper border of the temporalis muscle. The temporalis muscle was cut along the artery's axis in an inverted Y shape and dissected from the underlying bone, remaining attached to the linea temporalis superior. A bone flap with a diameter of 5 centimeters was created by connecting two burr holes at the limits of the artery's projection.

The dura mater was incised in an X-shaped pattern, carefully avoiding injury to the branches of the middle meningeal artery or coagulation. The thick arachnoid membrane over the cortex was dissected cautiously. The dura mater was reflected so its outer surface contacted the cortical surface beneath the skull. The posterior part of the temporalis muscle was placed in contact with the cortex, and the bone flap was replaced in its original position. The anterior part of the temporalis muscle was secured to the linea temporalis, stabilizing the bone flap. Skin closure was performed with standard techniques, and the area was covered with tissue. No additional bandaging was necessary.

Surgical Technique: EDAMS

The surgical technique of EDAMS is similar to that of EDMS, with the following differences.

First, a branch of the temporal artery was harvested during the skin incision. After the dural opening, arterial synangiosis with the harvested artery was carried out by attaching the adjacent cortex through the pia mater to the adventitia using an 11/0 silk suture.

Finally, bone flap fixation at closure allowed the patent artery to run beneath it after entering through the inferior burr hole and exiting through the superior one, ensuring the artery was not compressed at any point.

Surgical Technique: STA-MCA Bypass

Patients were prepared and positioned supine, with the head turned to the opposite side. An STA was dissected and distally clipped to a depth of 10 cm. A fish mouth incision of the distal edge of the artery is prepared for the anastomosis, a temporoparietal bone flap is harvested, the dura is opened, and the outer part of the Sylvian fissure is opened by arachnoid dissection. An M3 artery is selected, temporary clips are applied to the anastomosis area, and a terminal-lateral anastomosis is completed.

The heel, followed by the toe of the STA extremity, was stitched with two 10.0 nylon sutures; the first half-running suture was performed on the blind side, and the second was tightened to the first, after flushing clots with diluted heparin.

After the anastomosis was completed, the temporary clips were removed from the distal and proximal recipient arteries and finally from the STA, reestablishing the flow.

Dural closure starts from the frontal to the temporal side. The muscle stitches and bone flap fixation allow the STA to enter safely without compression.

Postoperative Period

Immediately after surgery, a neurological exam was performed, followed by a CT scan within 24 hours. If new neurological deficits or deterioration were detected, an MRI was then conducted. The patient received hyperhydration and transfusions if hemoglobin dropped below 12 g/dL.

Cilostazol (100 mg daily) was restarted on the second postoperative day. For the STA-MCA bypass, the patient was given intravenous aspirin three hours after the CTA in the ICU, along with preoperative diluted saline heparin and a calcium channel blocker.

Typically, the patient was discharged seven days after surgery and scheduled for a one-month follow-up, with evaluations every three months afterward. A postoperative angiogram using MRA with perfusion imaging was performed at six months. If financially feasible, angiography was conducted between 12 and 18 months postoperatively.

Literature Review Methodology

This systematic review examined pediatric patients (≤18 years) diagnosed with Moyamoya disease (MMD) or Moyamoya syndrome (MMS) in North Africa, including Algeria, Morocco, Tunisia, Libya, and Egypt. Its goal was to gather evidence on clinical presentation, imaging features, vessel involvement, treatment approaches, and follow-up outcomes for this population. A thorough search was conducted across international and regional databases, such as PubMed/MEDLINE, Scopus, Web of Science, Embase, and Google Scholar. To find reports not listed in these sources, additional regional databases like African Index Medicus, IMIST, and local neurosurgical society publications were also explored. The search covered the period from January 1980 to December 2025, using keywords and Boolean operators like "Moyamoya" AND "North Africa," "Moyamoya" AND (Algeria OR Morocco OR Tunisia OR Libya OR Egypt), "Pediatric" OR "child" OR "adolescent" AND "Moyamoya disease," and "Moyamoya syndrome" AND "Africa." References from relevant articles were manually checked to identify other pertinent studies.

The inclusion criteria comprised (1) pediatric patients (≤18 years) diagnosed with MMD or MMS, (2) cases from Algeria, Morocco, Tunisia, Libya, or Egypt, (3) study types including case reports, case series, retrospective cohorts, or prospective observational studies, and (4) sufficient clinical, imaging, management, or follow-up data. Studies focusing only on adults, lacking clear diagnosis confirmation, reviews, editorials, or those involving non-North African populations were excluded.

Two independent reviewers collected data with a standardized form covering demographics, clinical features, imaging, etiology, treatment, and outcomes. Disagreements were resolved through consensus. If datasets overlapped, the more detailed dataset was chosen.

Study quality was assessed using the Joanna Briggs Institute (JBI) Critical Appraisal Checklists, classifying studies as low, moderate, or high quality. Due to variations in study design and sample size, a descriptive synthesis was performed. Quantitative data were summarized as medians and ranges, while categorical data were presented as frequencies and proportions.

Results

Preoperative Assessment

The preoperative clinical and imaging evaluations for the nine patients are summarized in Table 1. The cohort included two males and seven females, aged between 1 and 15 years (average age 5 years). Eight patients exhibited neurological deficits, with one experiencing only headaches. Specific neurological impairments involved hemiplegia in two patients, hemiparesis in three, and upper-limb monoplegia in three. Ischemic strokes affected the frontal lobe in five patients, the parietal lobe in five, the occipital lobe in one, and the hemisphere in one. The preoperative pmRS scores were 5 in two patients, 4 in three, 3 in three, and 2 in one.

Table 1. Preoperative clinical and imaging features. N: number of patients; M: male; F: female; pmRS: pediatric modified Rankin scale; L: left; R: right; B: bilateral; ICA: internal carotid artery; PCA: posterior cerebral artery.

N	Age	Sex	Symptoms	pmRS	Stroke	Anterior	Posterior
1	3	M	Monoplegia of the right upper limb	4	L. fronto-parietal	B. ICA	R. PCA
2	1	F	Right paresis	4	L. hemisphere	B. ICA	B. PCA
3	3	F	Right hemiplegia, left upper limb paresis	5	R. frontal, L. parieto-occipital	R. ICA	-
4	5	F	Right hemiplegia	5	R. parietal	B. ICA	B. PCA
5	7	F	Left paresis, headaches	4	L. frontal, R. parietal	B. ICA	B. PCA
6	4	M	Monoplegia of the right upper limb	3	R. frontal	B. ICA	B. PCA
7	15	F	Headaches	2	R. frontal	R. ICA	-
8	5	F	Left hemiparesis	3	R. parietal	B. ICA	R. PCA
9	3	F	Monoplegia of the right upper limb	3	R. parietal	B. ICA	-

Vessel Involvement

This North African moyamoya cohort mainly shows anterior circulation involvement, with 100% (9/9) of patients affected on the right side and 77.7% (7/9) on the left. Posterior circulation changes are also common but less widespread, involving the right posterior cerebral artery in 66.6% (6/9) and the left in 44.4% (4/9). This pattern indicates a consistent right-side dominance in both anterior and posterior

regions. While anterior circulation stenosis is very frequent, posterior involvement occurs in nearly half to two-thirds of cases. As shown in Figure 1, involvement of the four brain vascular axes depends on disease progression: it starts with the anterior circulation, affecting the right internal carotid artery (ICA), then the left ICA. Over time, the lesions extend to the posterior circulation, beginning with the right posterior cerebral artery (PCA), followed by the left.

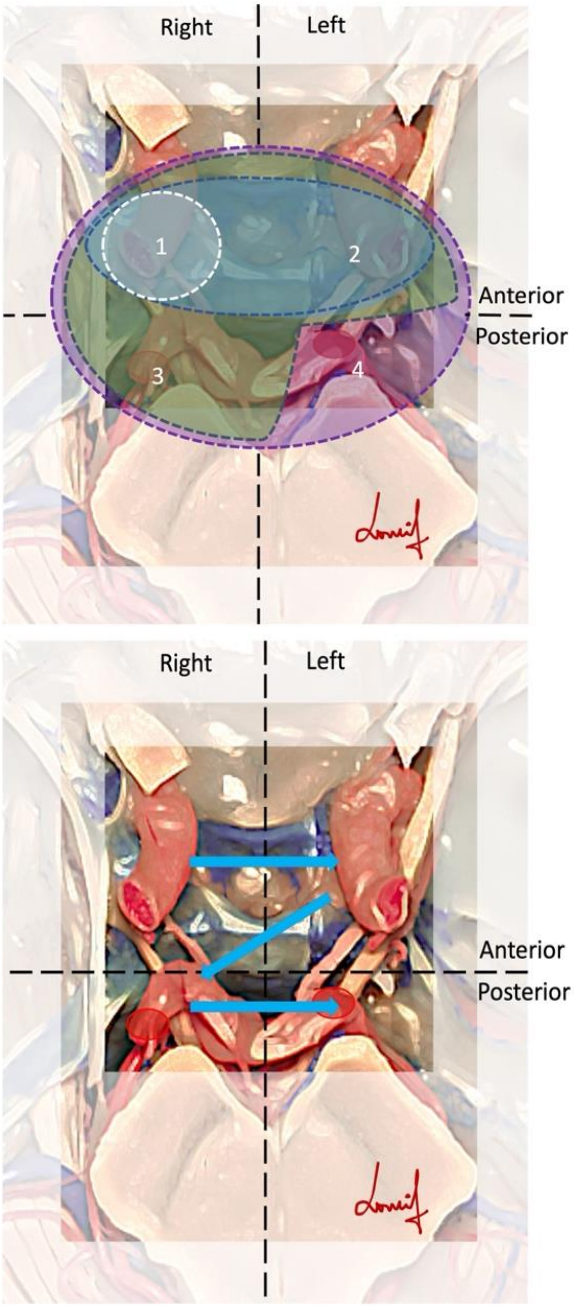

Figure 1. Involvement pattern with frequencies and dependency of the four vessel axes (A) and the "Z-shaped" progression of vessel involvement (B).

Postoperative Outcomes

Postoperative clinical, surgical, and imaging outcomes are summarized in Table 2. All patients received EDMS; four also underwent STA synangiosis (EDAMS), and one older child had an STA-MCA bypass. The choice of technique depended on age: EDMS was used in children under 4 years, EDAMS in those aged 5-8 years, and STA-MCA bypass was added selectively for patients over 8 years when anatomy allowed. Long-term pmRS scores improved in three patients with preoperative paresis, and no patient experienced neurological deterioration. Three cases had postoperative strokes, but all remained stroke-free after the early postoperative period and at 12 months. Follow-up ranged from 14 to 36 months, with a mean of 21 months.

Table 2. Postoperative surgical, clinical, and imaging outcomes. EDMS: encephalo-duro-myo-synangiosis; EDAMS: encephalo-duro-arterio-myo-synangiosis; Postop: postoperative; pmRS: pediatric modified Rankin scale; L: left; R: right; STA: superficial temporal artery; MCA: middle cerebral artery.

N	Surgical technique	pmRS Postop	Stroke Postop	Stroke 12mo	Follow-up (months)
1	EDMS	4	-	-	16
2	EDMS	2	L. frontal	-	18
3	EDMS	5	-	-	36
4	EDAMS	5	R. occipital	-	14
5	EDAMS	2	-	-	22
6	EDAMS	3	R. occipital	-	28
7	STA-MCA bypass + EDMS	2	-	-	19
8	EDAMS	2	-	-	20
9	EDMS	3	-	-	15

Illustrative Case

A 7-year-old girl presented with left hemiparesis and headache. MRI showed a left parietal ischemic stroke along with multiple white matter lesions. Angiography demonstrated bilateral carotid stenosis and a dense lenticulostriate network with a cigarette-puff pattern. Perfusion and diffusion imaging showed no mismatch, and comparison between the two parietal lobes indicated preserved perfusion on the right side, leading to the decision to operate only on the left.

The patient underwent EDAMS, an indirect revascularization procedure. The temporal artery and its branches were dissected, a bone flap was harvested, the dura was inverted, and the temporal artery was sutured to the pia mater with 11/0 stitches.

Postoperative angiography revealed increased cortical and meningeal neovascularization and a

regression of the moyamoya network compared to preoperative imaging.

Since the surgery, the patient has remained stroke-free, with only mild residual hemiparesis (3/5).

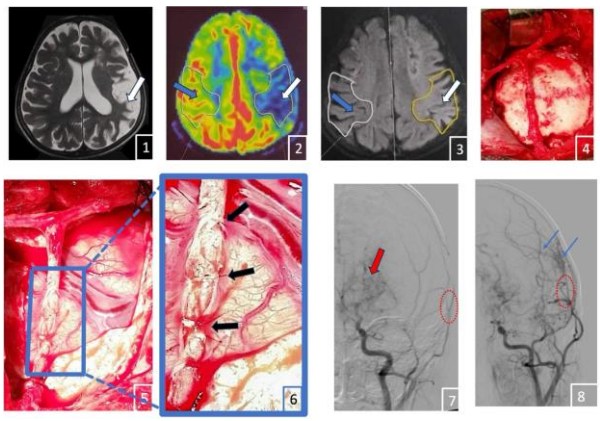

Figure 2. Imaging and surgical findings for the illustrative case. (1) Axial T2-weighted MRI scan showing an ischemic stroke in the left parietal lobe, along with multiple white matter lesions in the frontal lobe. (2) Perfusion MRI showing hypoperfusion in the left parietal area from the stroke; the opposite parietal lobe shows normal perfusion. (3) Axial diffusion-weighted MRI showing hypoperfusion in the left parietal lobe and normal perfusion on the right. (4) Perioperative image with the superficial temporal artery and its two branches dissected; the underlying bone flap was harvested but remains in place. (5) Surgical image showing the exposed frontotemporal cortex, with the temporal artery and branches on the cortex; sutures attached the artery to the pia mater, and the dura mater was reversed. (6) Enlarged view highlighting the temporal artery and branches on the cortex, with sutures attaching the artery to the pia mater. (7) Preoperative brain angiography depicting stenotic occlusion of the left carotid artery and a lenticulostriate network. (8) Postoperative angiography demonstrating revascularization of the left lobes with cortical neovascularization and developed meningeal arteries.

Discussion

Pediatric MMD is rare in North Africa, with sporadic cases reported in the literature.

In this study, we reported nine patients with MMD with a male-to-female sex ratio of 1:4.5. However, in the non-Asian population, the incidence of MMD in female patients is higher than that in male patients, but with the male-to-female ratio ranging from 1:1.8 to 1:2.2 [6-8]. By contrast, in China, the sex ratio was 1:1 [9]. This discrepancy could be explained by the small number of patients included in the present study.

In our study, clinical signs allowed us to differentiate between two age groups. We used a cutoff age of 5 years, determined by observations showing that children under this age exhibited more severe symptoms at onset, often indicating monoplegia or hemiplegia in the affected region. Conversely, children aged five and older typically showed milder symptoms, such as headaches and paresis. All cases involved ischemic strokes, with no reports of hemorrhagic strokes.

Vessel Involvement in MMD

MMD is a recognized clinical and pathological condition [7,10,11]. Traditionally, the condition involves progressive narrowing and blockage of the internal carotid arteries and their main branches, leading to the formation of fragile vessels at the brain's base. Most research has concentrated on the anterior circulation, with limited studies on the progression to the posterior circulation. However, cases involving the posterior circulation often show more severe symptoms and a poorer prognosis than those affecting the anterior circulation, especially in children. The development of vascular lesions across the four axes of the skull base remains poorly understood, with limited data on how they progress. It is generally believed that initial lesions usually occur in the anterior circulation, with carotid involvement being the key factor and the hallmark of MMD.

Genetic predisposition accounts for the steno-occlusive phenomenon in MMD. However, the previous involvement of the anterior circulation, especially the predominance of the right over the left, remains insufficiently explained. Additionally, involvement of the posterior circulation is characterized by involvement of the second segment of the posterior cerebral artery (P2) [12]. The different embryological origins of the first segment of the PCA (P1) and P2 can be explained as follows: P1 develops from the primitive vertebral basilar artery, while the fetal posterior communicating artery, a branch of the carotid artery, corresponds to the P2 segment of the PCA [13]. Its involvement in MMD lesions results from its progression from the carotid artery to the posterior communicating artery.

The primitive vertebral basilar artery system is rarely involved, and Tan et al. [12] suggested that the primitive basilar artery forms relatively late in brain development, which is why MMD primarily affects the brain's blood vessels. Unlike the embryonic development of the primitive internal carotid artery system, which occurs within the brain, the basilar

artery develops outside the brain, originating from the longitudinal dorsal nerve artery early in embryogenesis. In our series, the hypothesis progression pattern — from right to left and anterior to posterior — is unique and supported by statistical analysis. The tests show that anterior vessels (ICAs) are significantly more often involved than posterior vessels (PCAs, $p = 0.0009$). This is confirmed by a highly significant Chi-square test result ($p = 0.0009$). Conversely, the difference between right and left-sided involvement is not statistically significant ($p = 0.0748$), suggesting only a trend toward right-sided dominance that does not meet the 0.05 threshold. A larger, more representative patient population is needed to clarify the laterality of involvement.

Medical Treatment

After an ischemic stroke in patients with Moyamoya Disease (MMD), the main priority is to evaluate the need for possible surgical revascularization. Aspirin is administered as an additional measure for secondary prevention. In addition to its antiplatelet properties, aspirin effectively alleviates headache symptoms associated with MMD. Ando et al. [14] reported that cilostazol enhances cognitive function more effectively than other antiplatelet agents in adults diagnosed with MMD. Furthermore, a comprehensive analysis conducted by Chiba et al. [15] demonstrated that cerebral blood flow improved in patients with ischemic stroke who were not eligible for surgery and were treated with cilostazol, whereas no such improvement was observed in patients receiving alternative antiplatelet medications.

Cilostazol is a phosphodiesterase III inhibitor that produces antiplatelet, antioxidant, and vasodilatory effects by elevating the activity of intracellular cyclic adenosine monophosphate and endothelial nitric oxide synthase [16]. These differences from other antiplatelet drugs might enhance cerebral perfusion [15]. Cilostazol is regarded as a safe medication that has beneficial effects on hyperactivity in children with autism spectrum disorder and elevated hyperactivity levels [14]. The question still stands as to why it isn't recommended as a first-line treatment for children with MMD in the guidelines.

In addition to antiplatelet agents, other medications are used to prepare patients for surgery. Additionally, perioperative stress and pain can cause uncontrollable crying, which may affect blood carbon dioxide levels, potentially resulting in hypocapnia and increasing the risk of ischemia from vasoconstriction. Therefore, providing adequate analgesia is advisable [17].

Surgical Treatment

Managing MMD primarily involves surgical procedures, including both direct and indirect revascularization. These treatments aim to prevent the recurrence of ischemic and hemorrhagic strokes, as well as collateral vessel regression seen on angiography [10]. However, the number of revascularization procedures continues to rise, supported by studies showing improved long-term outcomes [4].

Duan et al. [18] investigated patients with MMD. It was indicated that surgically treated patients experienced a significantly lower recurrence of ischemic stroke compared to those managed conservatively. Consequently, the current consensus recommends that patients with ischemic or hemorrhagic MMD should receive surgical intervention [1,19].

Omote et al. [20] reported that EDAMS, with or without STA-MCA anastomosis, was more effective than simple EDMS in establishing collateral circulation in pediatric patients with MMD [21].

In young children, the diameters of the superficial temporal artery (STA) and middle cerebral artery (MCA) are smaller, making direct revascularization via bypass procedures particularly challenging. Veeravagu et al. noted that, for children under four, surgeons must tailor their intraoperative approach, choosing between direct and indirect revascularization based on the variable and small sizes of the STA and recipient vessels [22].

Regarding clinical presentation, surgical management classifies patients into two groups based on the STA diameter, which is often less than 1 mm in those under five years old. For this age group, revascularization using the EDMS technique is performed. In children over five, revascularization involving the temporal artery is recommended. Between ages 5 and 8, EDAMS is preferred. For children older than 8, a STA-MCA bypass may be added, but this decision is made preoperatively based on individual anatomical differences.

The procedure was performed bilaterally in the same session. However, a six-week interval is considered essential to prevent postoperative neurological complications, as reported by Deng et

al. [23] Three cases experienced immediate postoperative strokes, with occlusion of both anterior and posterior bilateral vessels. This accounts for the poor and unstable cerebral perfusion seen initially. However, neurological symptoms improved by the 12-month follow-up.

Conclusion

Moyamoya disease is rare in North Africa, but this series shows that affected patients, especially children, have distinct clinical and vascular features. The dominance of anterior circulation involvement and the statistically confirmed anterior-posterior progression offer new understanding of the disease's timeline in this population. The common misclassification of Twig-like MCA and Moyamoya syndrome in regional reports highlights the need for more accurate diagnosis. By presenting the largest regional cohort and reviewing published cases, this study provides vital epidemiological and radiological data and emphasizes the importance of increasing awareness and early diagnosis of Moyamoya disease in North African clinical practice.

Abbreviations

MMD: Moyamoya disease; MMS: Moyamoya syndrome; STA: superficial temporal artery; MCA: middle cerebral artery; pmRS: pediatric modified Rankin scale; L: left; R: right; B: bilateral; ICA: internal carotid artery; PCA: posterior cerebral artery; Pre-op: preoperative; Postop: postoperative; EDMS: encephalo-duro-myo-synangiosis; EDAMS: encephalo-duro-arterio-myo-synangiosis; CTA: computed tomography angiogram; RNF: ring finger protein; MRI: magnetic resonance imaging; MAP: mean arterial pressure; CT: computed tomography; M: male; F: female; Isch: ischemic; Hgic: hemorrhagic; TIAs: transient ischemic attacks.

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