ORIGINAL ARTICLE

Outcome of revascularization in moyamoya disease: Evaluation of a new angiographic scoring system

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ABSTRACT

Background: Moyamoya disease (MMD) is a chronic progressive cerebrovascular occlusive disease affecting commonly the anterior circle of Willis. Matushima grade inadequately reflects the angiographic changes postrevascularization procedure.

Aims: To analyze the clinical and angiographic outcome of revascularization procedures (direct [ST-middle cerebral artery (MCA) anastomosis] and indirect [encephalo-duro-arterio-myo-synangiosis (EDAMS)]) in MMD and validate a new angiographic scoring system.

Materials and Methods: Retrospective study included symptomatic patients of MMD who underwent revascularization; both indirect and combined methods between January 2002 and April 2012. Follow-up angiography was done after at least 3 months. We devised a novel scoring system the "angiographic outcome score" (AOS) including reformation of distal MCA and anterior cerebral artery, regression of basal moyamoya vessels, leptomeningeal collaterals and overall perfusion. AOS was applied to the angiograms independently by a neuroradiologist and a neurosurgeon that were blinded toward its preoperative or postoperative status.

Results: Totally 33 patients underwent 36 EDAMS and 4 combined procedures (EDAMS + ST-MCA bypass). The mean follow-up was 20 months. None had recurrent transient ischemic attack or fresh infarct. Postoperative AOS was significantly higher than preoperative AOS. The Spearman rho showed positive correlation between Matushima grade and postoperative AOS. Significant regression of basal moyamoya vessels and increase in number of loci of transdural collaterals was seen.

Conclusions: EDAMS is a simple yet effective method of revascularization in both pediatric as well as adult age groups. AOS is a simple, precise and easily reproducible scoring system, which reflects the favorable angiographic changes after revascularization.

Key words: Angiography, encephalo-duro-arterio-myo-synangiosis, moyamoya disease, revascularization

Introduction

Moyamoya disease (MMD) is a chronic progressive cerebrovascular occlusive disease, characterized by stenosis of bilateral supraclinoid internal carotid artery (ICA) and/or proximal anterior cerebral artery (ACA) and middle cerebral artery (MCA). The irregular vascular network of perforating collateral vessels at the base of brain close to the stenotic segments particularly in the region of lenticulostriate and thalamoperforate vessels, called the moyamoya vessels gives the characteristic angiographic appearance. Revascularization procedures in MMD include direct, indirect or combined procedures. Suzuki[1] staging [Table 1a] is currently most widely used for assessing disease severity and Matsushima[2] grade [Table 1b] describes the extent of perfusion on postoperative angiograms. While analyzing the angiography we felt the need to interpret the differences in the preoperative and postoperative angiograms more objectively. The attributes of an effective scoring system are simplicity, precision, universality, and reproducibility. No such scoring system exists in the literature. We devised a novel scoring system the "angiographic outcome score (AOS)," pertaining to the felt need, applied it to the present study group and validated it.

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Materials and Methods

The present study was conducted in a tertiary care hospital with the main objective of analyzing the clinical and angiographic outcome of revascularization in MMD and evaluation of a new angiographic scoring system. Clinical and imaging records were reviewed retrospectively.

Symptomatic patients of MMD: Who underwent revascularization; both indirect and combined methods, were included and who underwent procedures other than revascularization like embolization of aneurysms and patients lost to follow- up, were excluded from the study.

During a period of 10 years and 3 months (January 2002–April 2012), 33 patients were treated for MMD with 40 procedures on 40 hemispheres. The series included 17 pediatric (21 treated hemispheres) and 16 adult patients (19 treated hemispheres). The preoperative radiological assessment was done as per angiographic staging of MMD (Suzuki staging).[1] Surgical procedures were chosen on the basis of availability of an ideal donor and recipient vessels. Totally 36 EDAMS and 4 combined procedures (EDAMS + ST-MCA bypass) were done. Postoperative complications such as hematoma, seizures, worsening of previous neurological deficits, new neurological deficits or cerebrospinal fluid leak, if any, were documented.

Follow-up assessment was done to see any fresh episodes of stroke/transient ischemic attack (TIA), persistent headache, new neurological deficits and graded as per modified Rankin scale. Follow-up angiography was done after at least 3 months and assessed independently by 2 observers for the extent of revascularization. Clinical outcome was assessed on 32 (97%) patients available for follow-up. One patient from abroad was lost to follow-up. Follow-up angiograms of 20 operated hemispheres in 17 (52%) patients were available for comparison and angiographic outcome analysis.

The angiographic outcome score: A novel scoring system

The natural course of MMD is progressive occlusion of the anterior circle of Willis and the blood supply to the brain is by the interplay between the basal moyamoya collaterals, the transdural collaterals, and the leptomeningeal collaterals. Among these, the basal moyamoya vessels are the most unfavorable mode of compensation, as these thin, friable vessels are least likely to bear the hemodynamic stress and hence the causative agents for complications. Thus, the philosophy behind scoring the angiographic findings is to consider an uncompensated or unfavorably compensated disease at one end of the spectrum and a favorably compensated disease at the other end. Revascularization procedures are known to halt the natural course of the disease or redirect it in the direction of favorable compensation, which is observed as regression or no further intensification of basal moyamoya vessels, increase in the transdural collaterals and the leptomeningeal collaterals leading to increased perfusion.

Table 1a: Suzuki staging[1]

Stage	Angiographic findings
1	Narrowing of the internal carotid artery bifurcation without collaterals
2	Initiation of moyamoya collaterals
3	Intensification of moyamoya collaterals
4	Minimization of moyamoya collaterals
5	Reduction of moyamoya collaterals
6	Disappearance of moyamoya collaterals

Table 1b: Postoperative collateral grading on external carotid angiography (Matsushima and Inaba)[2]

Grade	Angiographic findings
A	Area perfused by the synangiosis is greater than 2/3 of the MCA territory
B	Area perfused by the synangiosis is between 1/3 and 2/3 of the MCA territory
C	Area perfused by the synangiosis is less than 1/3 of the MCA territory

MCA – Middle cerebral artery

Table 2: AOS (A new scoring system to analyze preoperative and postoperative angiograms)

	Score
Major vessels	
A1. MCA	
Poor	0
Fair	1
Good	2
A2. ACA	
Poor	0
Fair	1
Good	2
Collaterals	
B1.Basal moyamoya	
Intense	0
Moderate	1
Mild	2
B2.transdural collaterals (number of loci)	
1-2	1
≥3	2
B3.leptomeningeal collaterals	
Mild	0
Moderate	1
Intense	2
C. Perfusion	
<1/3rd	0
1/3rd-2/3rd	1
>2/3rd	2

The total score X=A (A1+A2) + B (B1+B2+B3) + C. Minimum score=1 and maximum score=12. AOS: Angiographic outcome score, MCA – Middle cerebral artery; ACA – Anterior cerebral artery

We grouped these disease-modifying features and assigned a score to each based on the following description. The total score was termed as the AOS [Table 2].

Major vessels

The MCA and ACA are reformed distally either by transdural collaterals, leptomeningeal collaterals or patent posterior communicating artery. The appearance on both anteroposterior (AP) and lateral projections were considered on ICA injection and graded as

- Poor: Barely visible, thread like vessels or absent reformation
- Fair: Intermediate between a and c
- Good: Almost resembling normal distal branches of ACA/MCA.

Collateral circulation

Basal moyamoya collaterals

- Mild: Minimal net like vessels, no puff of smoke appearance, corresponding to moyamoya vessels seen in Suzuki stage 1
- Moderate: Intermediate (between a and c)
- Intense: Extensive net like vessels with typical puff of smoke appearance corresponding to Suzuki stage 3.

Because intense moyamoya vessels is a unfavorable compensation it is given a low score as compared to mild basal moyamoya vessels which points towards a favorable compensation and begets a higher score.

Transdural collaterals

The transdural collaterals developed mostly in the distribution of branches of the middle meningeal artery. The various areas of transdural collateral development were categorized as ophthalmic, frontal, pterional, anterior and posterior parietal and occipital. The loci were observed under selective External Carotid Artery (ECA) injection. Hence, more the loci of transdural collaterals, the higher the score will be.

Leptomeningeal collaterals

The development of leptomeningeal collaterals is a favorable compensation and the more such vessels the higher the score.

- Mild: Limited to 1/3rd surface area on a lateral projection
- Moderate: Intermediate between a and c
- Intense: Extension up to ≥2/3rd surface area on lateral projection.

Perfusion

Scoring the perfusion was equivalent to Matushima grade but included a virtual summation of perfusion on both carotid and vertebral injection on lateral as well as AP projections. Increasing perfusion was given higher scores.

The total score X = A (A1 + A2) + B (B1 + B2 + B3) + C. the minimum score was 1 and the maximum score was 12. A lower score was suggestive of advanced disease and a higher score was suggestive of an early disease or a favorably compensated MMD. The preoperative and follow-up angiograms were given scores independently by a neuro-radiologist and a neurosurgeon, who were blinded toward the preoperative or postoperative status of the angiogram.

Statistical analysis

The inter-observer agreement on angiographic findings was studied by Kappa statistics. Wilcoxon signed-ranks tests were used for studying the differences between preoperative and postoperative angiographic outcome in the entire series. Mann–Whitney U test was used for studying the differences of angiographic outcome between both age groups. Spearman's rho was used for correlation studies. $P < 0.05$ was considered as significant. SPSS (IBM Corporation) 16 was used for statistical analysis.

Results

Clinical and angiographic presentation

Ischemic symptoms were present in 82% patients (all pediatric and 62% adult patients) with recurrent TIAs in 6%, (all children). Hemorrhagic presentation was seen in 18%, only in adult patients, with intra-ventricular hemorrhage in three patients [Table 3a].

Hemiparesis (78%) followed by the seizure (18% of total and 30% of pediatric patients) was the presenting feature. Headache was present initially in 24% patients. Painless, progressive visual deterioration and delayed development were seen in 18% and 6% of pediatric patients respectively.

Twenty-four preoperative angiograms (48 hemispheres: 22 adult and 26 pediatric) were available for analysis. Disease

Table 3a: Patient profile

	(%)
Age (years)	
Mean	20
Range	3-45
Sex	
Male	18 (55)
Female	15 (45)
Pediatrics	17 (52)
Adults	16 (48)
Clinical presentation	
Ischemic	27 (82)
Hemorrhagic	6 (18)
Surgical procedure	
Total number of patients undergoing revascularization	33
Total number of EDAMS	36
Total number of STA/MCA bypass (combined procedures)	4
Total number of B/L EDAMS	07 (18)

MCA – Middle cerebral artery; EDAMS – Encephalo-duro-arterio-myo-synangiosis; STA – Superficial temporal artery

severity of Suzuki stage ≥4 was present in 75% hemispheres at initial presentation. Asymmetry of Suzuki stage of both the hemispheres was seen in 29% patients. Posterior cerebral artery (PCA) involvement was seen in 25% hemispheres and common in pediatric 42% patients [Table 3b].

Revascularization procedures and complications

Forty revascularization procedures were done in 33 patients: 20 EDAMS were done in pediatrics and 16 in adults, whereas 3 combined procedures were done in adults and 1 in a pediatric patient.

Postoperative complications were seen in 15% patients and most were minor. Neurological deterioration was seen in only one adult patient with the development of hemiplegia and increase in the size of infarct on the operated side. This was attributable to perioperative dehydration due to multiple episodes of vomiting. The patient was managed conservatively and subsequently improved. Two patients had seizures postoperatively, controlled with an increase in antiepileptic drug dose. Wound infection and chronic subdural hematoma developed in one patient each, which required wound debridement and evacuation of the subdural hematoma, respectively.

Clinical outcome

The mean follow-up was 20 months. Clinically, 81% of all patients had improvement in hemiparesis and a modified rankin scale of 0 or 1 at last follow-up. None had recurrent TIA or fresh infarct till last follow-up. One adult patient, who had presented with intra-ventricular hemorrhage and was operated by combined procedure (superficial temporal artery [STA]/MCA bypass and EDAMS), developed recurrent episode of intra-ventricular hemorrhage at 120 months of follow-up for which he was treated conservatively. Persistent headache was present in 16% patients [Table 4].

Angiographic outcome: Angiographic outcome score-evaluation of a new scoring system

Angiographic follow-up was available for 20 operated hemispheres (9 adults and 11 pediatric) with 17 EDAMS and 3-combined procedures [Table 3b]. The inter-observer agreement on angiographic findings was good ($\kappa = 0.620$) for major vessel reformation score, moderate ($\kappa = 0.548$) for collateral circulation and very good ($\kappa = 0.821$) for perfusion score. Matushima grade A and B perfusion was seen in 60% (12 of 20) and 30% (6 of 20) of the operated hemispheres respectively [Table 5]. The postoperative AOS was higher, same and lower in 85%, 15% and 0% hemispheres respectively, and this difference was statistically significant ($P = 0.000$) [Figure 1]. The Spearman rho showed a positive correlation (0.507) between Matushima grade and postoperative AOS, which was statistically significant ($P = 0.022$). Thus, a higher postoperative AOS over the preoperative score reflected favorably compensated MMD.

Table 3b: Preoperative and postoperative angiographic features

Angiographic feature	*n* (%)			
	Preoperative		Postoperative	
Suzuki stage				
1-3	3 (15)		2 (10)	
4 and above	17 (85)		18 (90)	
Major vessels	MCA	ACA	MCA	ACA
Poor	8 (40)	9 (45)	3 (15)	5 (25)
Fair	11 (55)	9 (45)	5 (25)	9 (45)
Good	1 (5)	2 (10)	12 (60)	6 (30)
Basal moyamoya vessels				
Intense	11 (55)		1 (5)	
Moderate	6 (30)		7 (35)	
Mild	3 (15)		12 (60)	
Leptomeningeal collaterals				
Mild	6 (30)		4 (20)	
Moderate	8 (40)		11 (55)	
Intense	5 (25)		5 (25)	
Transdural collaterals				
1-2	6 (30)		3 (15)	
≥3	14 (70)		17 (85)	
Perfusion				
<1/3rd	8 (40)		2 (10)	
1/3rd-2/3rd	9 (45)		6 (30)	
>2/3rd	3 (15)		12 (60)	

ACA – Anterior cerebral artery; MCA – Middle cerebral artery

Table 4: Outcome

Clinical outcome	
Mean follow-up	20 months (range 3-121 months)
Clinical follow-up	32/33 (97%)
Improvement in hemiparesis	26/32 (81%)
Recurrent TIA/infarct	0
Recurrent hemorrhage	1/32 (3%)
Persistent headache	5 (16%)
Angiographic outcome	
Operated hemispheres with angiographic follow-up	20
Matushima grade (%)	
A	12/20 (60)
B	6/20 (30)
Postoperative AOS > preoperative AOS	17/20 (85)
Regression of basal moyamoya vessels	11/20 (55)
Increase in loci of transdural collaterals	16/20 (80)

AOS – Angiographic outcome score; TIA – Transient ischemic attack

In the adults, the postoperative AOS was higher, same and lower than the preoperative score in 67%, 33% and 0% hemispheres respectively, and the difference was statistically significant ($P = 0.027$). The postoperative AOS was higher than the preoperative score in all-pediatric patients, and the difference was also statistically significant ($P = 0.003$). Thus, the AOS reflects the angiographic changes of revascularization procedures in both adults and pediatrics [Table 4].

Thus, AOS is a novel scoring system that provides a tool to comment more precisely on the extent of favorable collaterals established, perfusion achieved and hence the success of revascularization procedure. It is simple to use, precise, applicable to all age groups and easily reproducible.

Discussion

The first description of MMD came in 1957 by Takeuchi and Shimizu as "hypo genesis of bilateral internal carotid arteries".[3] Suzuki and Takaku introduced the term "moyamoya" in 1969 to characterize the angiographic appearance of the condition.[4] The international classification of disease has adopted the term moyamoya for the description of this clinical entity.[1]

Reported annual incidence and prevalence rate in Japan are 0.35 and 3.16/100,000 respectively with age at onset having two peaks, one at 5 years, and another at 40 years.[5] The disease is more common in females as compared to males (1.8:1).[5] The ischemic type predominates in childhood (69% of cases in patients <10 years old).[6] The hemorrhagic type is more characteristic of adults. It is extremely rare for children to present with hemorrhage (2.8%).[7]

In children, headache is a frequent presenting symptom, which may persist even after successful revascularization.[8] Epilepsy and involuntary movements are also commonly seen.[9,10]

At diagnosis, adults are usually at a more advanced stage than children, who progress more rapidly, especially patients younger than 2 years of age.[11,12] The symptomatic progression differs markedly in the surgically revascularized patients as compared to those untreated (2.6% vs. 66%) over a 5 years period.[13-16] Neurological status at the time of treatment is the single most important predictor of outcome, hence early diagnosis and treatment is of paramount importance.[9,17]

Digital subtraction angiography (DSA) is the gold standard for the diagnosis of MMD.[1,16] Classically bilateral supraclinoid ICA narrowing with narrowing of the MCA and ACA origins is seen with the involvement of PCA in about 25% of cases.[18] The basal collaterals appear as a net of vessels giving the appearance of a puff of smoke.

Surgical treatment started with cervical carotid sympathectomy by Suzuki *et al.* way back in 1973.[19] Revascularization for MMD can be accomplished by direct or indirect procedures. Direct methods include creating actual bypasses, like a STA-MCA bypass [Figure 2].[20-22] Indirect methods most commonly used are encephalo-duro-arterio-synangiosis (EDAS),[23] encephalo-myo-synangiosis[24] and various combinations of the two. The other procedures include multiple burr holes, dural inversion, galeal apposition, and omental transplants.[21,25]

Table 5: Angiographic outcome in adults and pediatrics (comparison of follow-up and preoperative angiograms)

	Adults (*n* (%))		Pediatrics (*n* (%))		*P* (adults vs. pediatrics)
Matushima grade					
A	4 (44)		8 (73)		0.331
B	4 (44)		2 (18)		
C	1 (12)		1 (9)		
Moyamoya vessels					
Regression	5 (55)	*P*=0.038	6 (56)	*P*=0.023	0.412
Same	4 (45)		5 (44)		
Intensification	0		0		
Transdural collaterals					
Increase	8 (88)	*P*=0.010	8 (73)	*P*=0.013	0.503
Same	1 (12)		2 (18)		
Decrease	0		1 (9)		
AOS					
Increase	6 (67)	*P*=0.027	11 (100)	*P*=0.003	0.010
Same	3 (33)		0		
Decrease	0		0		

AOS – Angiographic outcome score

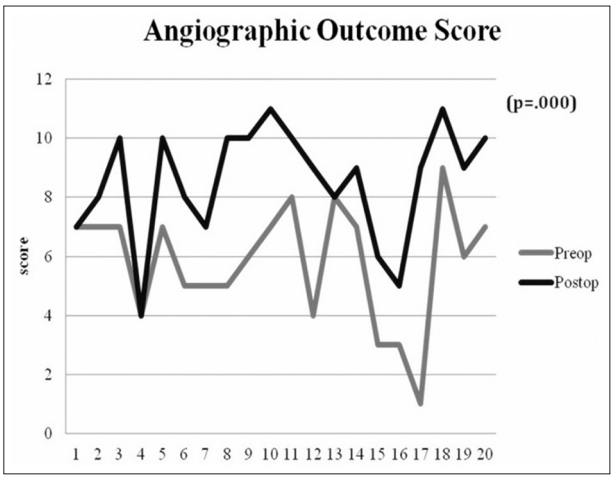

Figure 1: Preoperative and postoperative angiographic outcome score

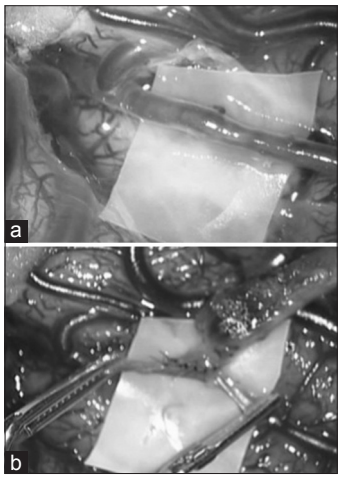

Figure 2: (a and b) Intraoperative image during superficial temporal artery-middle cerebral artery (MCA) bypass. a. Isolation of distal MCA branch. b. Completed anastomosis

Encephalo-duro-arterio-myo-synangiosis, in which the STA and the muscle flap are approximated to the surface of the brain and sutured to the dural edge was proposed and developed in 1984 by Kinugasa *et al.*[26] and found extensive collaterals in 28 sides operated in 17 patients by EDAMS when compared to patients operated earlier by EDAS.

Follow-up DSA to assess the development of collaterals after direct or indirect bypass surgery, should be done after a period of 3 months as time taken to develop collaterals about 3–4 months after indirect bypass surgery.[27] Moyamoya vessels start disappearing about 1-month after combined bypass surgery and the STA and middle meningeal artery increase in their diameters, which can be appreciated about 3 months after surgery.[28]

The revascularization procedures are known to regress or halt the intensification of basal moyamoya vessels, increase the trans-dural collaterals and the lepto-meningeal collaterals leading to increased perfusion and favorable hemodynamic compensation[23,24,26] clinically resulting in preventing further episodes of stroke in patients with MMD.[29]

In a study by Robertson *et al.*,[30] well developed transdural and transpial collaterals were seen after synangiosis in 84% of the surgically treated hemispheres in pediatrics at a mean follow-up of 12 months. Kim *et al.*[31] compared the angiographic and clinical differences of EDAS and EDAMS in 12 and 5 children respectively and concluded that most extensive collaterals were formed in the later. EDAMS was the indirect method of revascularization done in all patients in the present series. Nissim *et al.*[32] demonstrated on average 85% revascularization rate over 2 years after EDAMS. Kim *et al.*[33] concluded EDAMS with or without STA-MCA bypass had significant angiographic revascularization as compared to EDAS in pediatric patients with MMD.

In our series, 85% of the operated hemispheres showed improvement in collateral circulation with increase in perfusion over a mean follow-up of 20 months and a statistically significant rise in the postoperative AOS over the preoperative score ($P = 0.000$). The basal moyamoya vessels regressed, remained same and intensified on follow-up angiograms as compared to preoperative angiograms in 55%, 45% and 0% operated hemispheres respectively and the difference was statistically significant ($P = 0.002$). The number of loci of transdural collaterals on follow-up angiograms increased, remained same and decreased in 80%, 15% and 5% of operated hemispheres as compared to preoperative angiograms, which was also statistically significant ($P = 0.000$). Thus successful EDAMS resulted in an increase in number of transdural collaterals and regression of basal moyamoya vessels resulting in overall favorable hemodynamic compensation [Figure 3].

Direct revascularization is suggested to be advantageous in adults in view of adequate size of donor and recipient vessels with immediate restoration of cerebral blood flow; however, the complications are reported in form of cerebral hyperperfusion, infarction and intracerebral hemorrhage.[34,35] The disadvantages in children are the small size of donor and recipient vessels, the risk of compromising already existing transdural collaterals, and the increased risk of perioperative infarct in case of graft thrombosis or stasis of blood flow across the graft.[36,37]

In the present series, 3 hemispheres (2 adults, 1 pediatric) of combined procedure (STA-MCA bypass on the same side of EDAMS) were available for angiographic follow-up. There was no statistically significant difference in the angiographic outcome, in terms of the regression of moyamoya vessels, increase in number of transdural collaterals and overall perfusion ($P > 0.05$). The reformation of distal MCA branches in two adult patients remained same on the follow-up angiograms [Figure 4] as compared to poor visualization in the pediatric patient as compared to preoperative angiograms. The less number of patients can explain this difference with the combined procedure at our center.

Comparison between adults and pediatrics

In a study by Houkin *et al.*[38] in patients of MMD presenting with intra-cerebral haemorrhage, 100% pediatric patients undergoing EDAMS had a good outcome as compared to 68% with direct bypass; whereas in adults, 100% patients treated with direct bypass had a good outcome as compared to 38% patients treated with EDAMS.

Dusick *et al.*[39] showed good response to EDAS in 100% pediatrics and 90% adults, where the good response was defined as the absence of new ischemic events after 1-month postoperatively. However, the percent increase in vessel diameter of the middle meningeal artery was seen in 71% pediatric and 39% adults though the results were statistically insignificant.

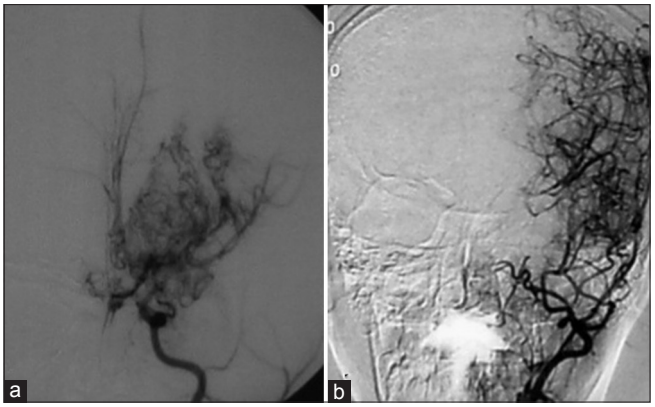

Figure 3: Preoperative angiogram a. Internal carotid artery injection: Anteroposterior (AP) projection and follow-up angiogram after encephalo-duro-arterio-myo-synangiosis b. External Carotid Artery injection: AP projection

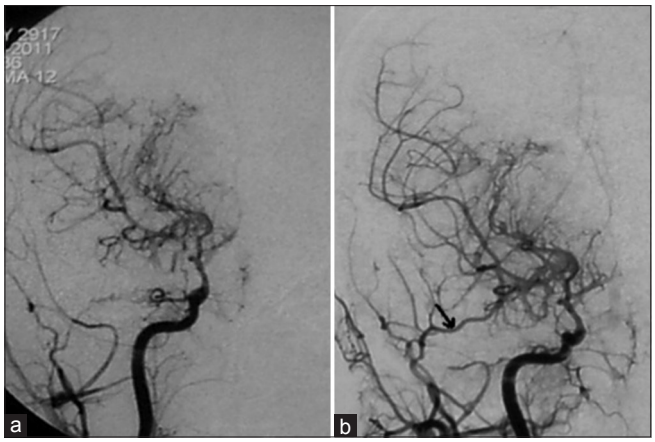

Figure 4: Preoperative a. and postoperative b. angiogram after superficial temporal artery-middle cerebral artery bypass showing the patent bypass

In a recent series by Lee *et al.*[40] (150 procedures in 106 patients), extent of revascularization and decrease in moyamoya vessels in adult patients was significantly more in patients treated with direct bypass or combined procedure when compared to indirect methods only.

In the present series, 81% patients had improvement in hemiparesis with modified rankin scale of 0–1 at last follow-up. No patients developed new onset ischemic episodes during follow-up. Matushima grade A perfusion was seen in 73% in pediatric and 44% of adult hemispheres, whereas Matushima grade B perfusion was seen in 18% of pediatric and 44% of adult hemispheres, but the differences were not statistically significant ($P = 0.331$). However, pediatric patients had a higher postoperative AOS as compared to adults, which was statistically significant ($P = 0.010$) by Mann–Whitney U test. The discrepancy from Matushima grade can be explained by the fact that, Matushima grade is based only on the proportionate perfusion of MCA territory on ECA injection whereas the AOS includes-reformation of distal MCA and ACA, regression of basal moyamoya vessels, leptomeningeal collaterals and overall perfusion. Thus, EDAMS was successful as the only revascularization procedure in both adults and children, though the angiographic outcome was more pronounced in children [Table 5].

Conclusions

Encephalo-duro-arterio-myo-synangiosis is a simple yet effective method of revascularization in both pediatric and adult age groups with MMD. Most of the patients treated with EDAMS had a good clinical outcome. AOS is a simple, precise and easily reproducible scoring system, which reflects the favorable angiographic changes after EDAMS.

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