Long-Term Surgical Outcome of Indirect Bypass Surgery in Young Children With Moyamoya Disease

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BACKGROUND: The prognosis of moyamoya disease (MMD) in young children (younger than 4 years) is worse than that of older adults. The effectiveness of surgery is still inconclusive.

OBJECTIVE: To evaluate long-term outcomes after indirect bypass in young children with MMD.

METHODS: A total of 1417 MMD children underwent indirect bypass from August 1988 to October 2020. This study included 135 patients who were younger than 4 years at the time of surgery. The clinical features and surgical outcomes of these patients were assessed. We analyzed the long-term outcome of 102 children who were followed up for more than 5 years (mean: 18.8 years, range: 5-27.3 years). Cross-sectional analysis was performed to evaluate overall outcomes based on the Lansky Play Performance Scale (LPS). The annual risk of symptomatic stroke after surgery was calculated with a person-year method, and the event-free survival rate was evaluated using the Kaplan-Meier method.

RESULTS: The overall clinical outcome was favorable (LPS ≥ 80) in 88% of the patients. The overall postoperative adverse event rate was 15%, including 1 death. At the last follow-up, 86% of patients who had seizures at diagnosis were seizure-free. During the follow-up, there were 3 symptomatic infarctions on the operated hemisphere (postoperative 3, 3, and 10 months each). There was no hemorrhagic event. The annual infarction rate was 0.16% per person-year. The 20-year event-free survival rates for symptomatic infarction were 97%.

CONCLUSION: Indirect bypass could provide a satisfactory long-term outcome and prevent recurrent stroke in young children with MMD.

KEY WORDS: Children, Infant, Moyamoya disease, Indirect revascularization, Stroke, Outcomes

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Moyamoya disease (MMD) is a chronic progressive cerebrovascular occlusive disease characterized by stenosis of distal internal carotid arteries with the development of a hazy network of collateral circulation.[1] The clinical presentation of MMD is repeated transient ischemic attacks (TIAs).[2] The benefit of indirect bypass for the ischemic type of pediatric MMD has been established.[3-7]

ABBREVIATIONS: EDAS, encephaloduroarteriosynangiosis; **EDH,** epidural hematoma; **EGPS,** encephalogaleoperiostealsynangiosis; **K-WISC-IV,** Korean-Wechsler Intelligence Scale for Children-IV; **LPS,** Lansky Play Performance Scale; **MMD,** Moyamoya disease; **OA,** occipital artery; **OP,** operation; **PCA,** posterior cerebral artery; **SDH,** subdural hematoma; **STA,** superficial temporal artery; **TIAs,** transient ischemic attacks.

Young-age MMD (younger than 4 years) is rare and has been considered the most severe and difficult to treat among subgroups of patients with MMD.[8,9] Their clinical course is dynamic and fast, leading to recurrent stroke and poor clinical outcomes.[10] Furthermore, infants are also at higher risk of anesthesia than older adults. Therefore, surgery for young patients with MMD proceeds with several risks and concerns.

We previously reported the clinical features and short-term surgical outcomes of patients with MMD younger than 3 years.[9] There are only a few studies dealing with the occurrence of

cerebral infarction immediately after surgery and during long-term follow-up in this group.[8-10] Therefore, the clinical study on the preventive effect of indirect bypass surgery on late-onset stroke is still needed. Here, we report the long-term surgical outcomes in the largest number of young patients with MMD treated with a uniform treatment strategy at a single institute. In this cohort, patients younger than 4 years were enrolled instead of 3 years.

METHODS

Patient Population

We reviewed all medical records of pediatric patients with MMD who underwent indirect bypass surgery between August 1988 and October 2020. MMD was confirmed through cerebral angiography or magnetic resonance imaging (MRI). We performed indirect bypass on 1417 pediatric patients during the study period. Young-age MMD was defined as a patient younger than 4 years at the time of surgery (N = 149). Unilateral MMD (N = 16) was included in this study. We excluded moyamoya syndrome (N = 14), a moyamoya-like angiopathy that occurs in association with various systemic diseases and conditions, such as neurofibromatosis type 1 and Down syndrome.

We reviewed 135 patients to evaluate the clinical features. To investigate long-term stroke incidence after surgery, we additionally analyzed 102 patients who were followed up for at least 5 years postoperatively (Figure 1). The mean follow-up duration was 18.8 years (range, 5-27.3 years). Patient consent was waived under Institutional Review Board approval (2105-106-1219) for this retrospective study.

Operative Technique

Surgery was performed in 2 stages, except for patients with unilateral MMD, which is usually in the symptomatic and the more hemodynamically affected hemisphere first, followed by the other side. On the other hand, in cases of symptomatic infarctions, we operated on the healthy hemisphere first to preserve at least 1 whole hemisphere. In the case of symptomatic infarction, surgery was performed after stabilization for approximately 2 weeks.

Encephaloduroarteriosynangiosis (EDAS) using the superficial temporal artery (STA) was performed for middle cerebral artery (MCA) area, in most cases. For anterior cerebral artery (ACA) territories, bifrontal craniotomy encephalogaleoperiostealsynangiosis (EGPS) or multiple burr hole EGPS was performed (N = 100).[11-13] In the posterior cerebral artery (PCA) territory, EDAS using the occipital artery (OA) or multiple burr hole EPGS was performed (N = 66).[14]

"Double craniotomies such as unilateral STA EDAS combined with bifrontal craniotomy EGPS, unilateral STA EDAS combined with OA EDAS, and bilateral OA EDAS had higher rates of symptomatic infarction compared with other procedures. A single craniotomy with multiple burr hole EGPS was relatively safer than 2 craniotomies."[15] Recently, we preferred to perform multiple burr hole EGPS for ACA and PCA territories.

Postoperative infarctions included an increased extent of previous infarction or a new-onset infarction accompanying new neurological symptoms. Postoperative adverse events were defined as infarctions or hemorrhages within 2 weeks after the operation.[6]

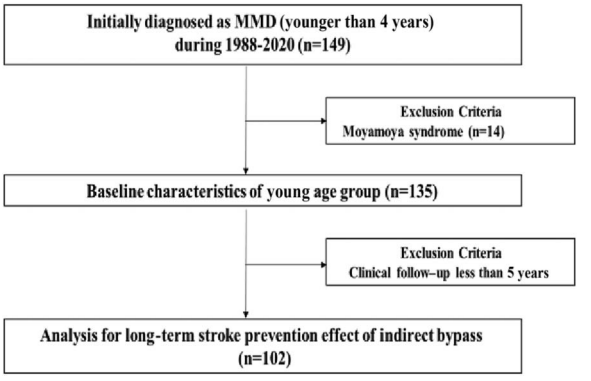

FIGURE 1. *Study subjects. From 1988 to 2020, 149 young patients with MMD (younger than 4 years at the time of surgery) underwent indirect bypass surgery. Excluding patients with moyamoya syndrome, 135 young patients with MMD were included. In addition, to assess the long-term stroke prevention effect, we analyzed 102 children who were followed up for more than 5 years. MMD, moyamoya disease.*

Postoperative Follow-up

Two weeks after surgery, the patient visited the outpatient clinic to observe the postoperative course. MRI, magnetic resonance angiography, and perfusion MRI were performed at 6 months, 1, 2, 4, and 6 years, and outpatient visits without MRI were performed at 8, 10 years, and thereafter at 5-year intervals. Patients were instructed to visit the emergency department or outpatient clinic if they had symptoms. Neurocognitive function was tested 2 and 4 years after the last surgery.

Cross-Sectional Analysis

After discharge, the clinical outcome was ascertained through clinical visits and imaging follow-up. Telephone interviews with patients and parents were also conducted for the most recent status check. The outcome was analyzed with 102 patients followed for more than 5 years. The overall clinical outcomes were evaluated using the Lansky Play Performance Scale (LPS) to measure neurological functional status. This scale classifies patients according to their functional impairment. The score ranges from 0 to 100. The lower the score, the worse their functional status. The favorable outcome was defined as LPS being 80 or more. The definition of LPS 80 means that the patients restrict strenuous play and tires more easily, otherwise active.

Seizure-free was defined as a patient experiencing no seizures for the previous 12 months or longer.

Intellectual status was reviewed based on Korean-Wechsler Intelligence Scale for Children-IV (K-WISC-IV) or descriptions on medical records or telephone interview. According to K-WISC-IV, a converted score of 90 to 109 is classified as average, 80 to 89 as below average, 70 to 79 as borderline, and below 69 as mental retardation.

In addition, we analyzed whether there was a difference between the favorable and unfavorable outcome groups in the age at operation, initial symptomatic infarction, subsequent preoperative infarction, initial LPS, postoperative complications, and PCA involvement. Univariate and multivariate logistic regression were performed to identify factors that showed significant differences between the 2 groups associated with unfavorable outcomes.

Longitudinal Analysis

A new stroke event was defined as any cerebral ischemia or hemorrhage with neurological symptoms that occurred more than 30 days after surgery.

Statistical Analysis

Continuous variables are expressed as the mean ± SD. Paired *t*-tests were performed to compare clinical status of patients before and after operation. Pearson correlation tests were used to determine the correlation between clinical features and poor functional outcomes. The annual risk of symptomatic infarction or hemorrhage in the operated hemispheres was calculated with a person-year method. The Kaplan-Meier method was used for stroke-free survival analysis. End points are stroke events or last follow-up (censored), and the criterion for statistical significance is $P < .05$.

RESULTS

Demographics and Clinical Presentation (Table 1)

A total of 135 young patients with MMD were included, consisting of 69 girls and 66 boys. The mean age at operation was 2.4 (range, 0.8-3.9) years. All the initial presenting types were ischemic. Among these, 113 (83%) patients had infarctions confirmed by preoperative MRI. Symptomatic infarction that included MCA, ACA, PCA territorial, or larger infarctions beyond the arterial territory occurred in 84 patients (62%). Others were border zone or small asymptomatic infarction. Only 1 patient had hemorrhage (1%). Hemorrhagic transformation of the infarcted area was observed. A total of 66 (49%) patients visited the hospital with seizures as the main symptom. Among them, 55 patients were associated with infarction.

Ten (8%) patients suffered from subsequent preoperative infarctions, suggesting rapid disease progression. A familial history of MMD existed in 28 (21%) patients. Unilateral MMD was present in 16 (12%) patients at presentation, and 8 progressed to bilateral disease during follow-up. With 1 exception (50 months), progression to bilateral disease was less than a year (2-11 months).

Interestingly, PCA involvement at presentation was as high as 27% of patients (N = 36). During follow-up, PCA involvement occurred in 55 patients (progression in contralateral PCA in 11 patients and newly developed PCA involvement in 44 patients).

TABLE 1. Summary of Demographics and Clinical Presentation (N = 135)

Baseline characteristics		No. of cases or value
Sex	Girl:boy	69:66
Age		2.4 (0.8-3.9)
Initial presentation	TIA	12
	Infarction	113 (83%)
	Symptomatic infarction	84 (62%)
	Seizure	66 (49%)
	Involuntary movement	3
	Hemorrhage	1 (with infarction)
Subsequent infarction	Y	10 (7%)
Family history	Y	28 (21%)
Unilateral	At presentation	16 (12%)
	Progression to bilateral	8
PCA involvement	At presentation	36 (27%)
	Progression	55 (41%)
	Contralateral side	11
	Newly developed	44
Extracranial involvement	Renal artery involvement	7 (5%)
	Pulmonary artery involvement	2 (2%)

PCA, posterior cerebral artery; TIA, transient ischemic attack; Y, yes.

Eventually, 80 (N = 36 + 44) patients (60%) had PCA involvement.

Renal artery involvement at presentation was observed in 7 patients. Three additional patients were diagnosed with renovascular hypertension during the follow-up period. Pulmonary artery involvement was rare (N = 2).

Surgery (Table 2) and Postoperative Complication (Table 3)

A total of 298 operations were conducted. Various combinations of surgeries were performed according to the patient's hemodynamic status at the time of surgery. STA EDAS was performed in 132 cases.

STA EDAS and bifrontal craniotomy EGPS (N = 68) or multiple burr hole EGPS (N = 29) were simultaneously performed in 97 cases. Surgery for the PCA area was performed with OA EDAS (N = 16) or multiple burr hole EGPS (N = 24) in combination with STA EDAS if PCA was initially involved. If PCA progressed during follow-up, additional OA EDAS multiple burr holes were performed (N = 42).

The average number of operations per person was 2.2 (range: 1-4). The overall postoperative adverse event rate was 15% per operation and 32% per patient. One patient died from infarction after surgery. Therefore, the overall mortality rate was 0.3% per operation and 0.7% per patient.

Infarction was the dominant complication, occurring in 25 cases (8% per operation and 16% per patient). The other complications were epidural and subdural hematoma in 14 cases (5% per OP and 10% per patient). Five of them required surgical removal. There was no parenchymal hemorrhage. Eight cases underwent revision surgery because of a wound problem.

TABLE 2. Types of Surgery

Types of surgery	Cases
STA EDAS	132
Bifrontal craniotomy EGPS	2
Bifrontal multiple burr hole EGPS	1
Unilateral OA EDAS	13
Unilateral occipital multiple burr hole EGPS	15
Bilateral OA EDAS	3
Bilateral occipital multiple burr hole EGPS	11
STA EDAS + bifrontal craniotomy EGPS	68
STA EDAS + bifrontal multiple burr hole EGPS	29
STA EDAS + OA EDAS	16
STA EDAS + OA multiple burr hole EGPS	8
Total	298

EDAS, encephaloduroarteriosynangiosis; EGPS, encephalogaleoperiostealsynangiosis; STA, superficial temporal artery; OA, occipital arter.

TABLE 3. Surgery and Its Complications (N = 135)

Parameters	Number of cases or value
No. of patients	135
No. of operations	298
Average operation per person	2.2 (1-4)
Duration between 2 staged operation (115 patients)	4 wk (1-28)
Postoperative adverse event	
Overall	45[a] (15% per OP, 32% per patient)
Death	1[b] (0.3% per OP, 0.7% per patient)
Infarction	25 (8% per OP, 16% per patient)
EDH and SDH	14 (5% per OP, 10% per patient)
Revision operation	5
Wound problem	8 (3% per OP, 6% per patient)

EDH, epidural hematoma; OP, operation; SDH, subdural hematoma.
[a]Two cases of infarction were simultaneously occurred with extra-axial hemorrhage.
[b]Death was due to massive infarction after surgery.

Cross-sectional analysis included overall clinical outcome, seizure-free rate, and intellectual status (Table 4)

These are the results of 102 patients followed up for more than 5 years. The overall clinical outcome was favorable in 90 (88%) patients based on their final LPS (≥80). The mean final LPS (90.6 ±

TABLE 4. Cross-Sectional Analysis: Overall Clinical Outcome, Seizure-Free Rate, and Intellectual Status (N = 102)

Parameters	Number of cases or value
Favorable outcome (LPS ≥ 80)	90 (88%)
Seizure-free rate	38/44 (86%)
Intellectual status (N = 84)	
K-WISC-IV (Score ≥ 70)	20/42 (48%)
Norm confirmed by interview or medical records	31/42 (74%)

K-WISC-IV, Korean-Wechsler Intelligence Scale for Children-IV; LPS, Lansky Play Performance Scale.

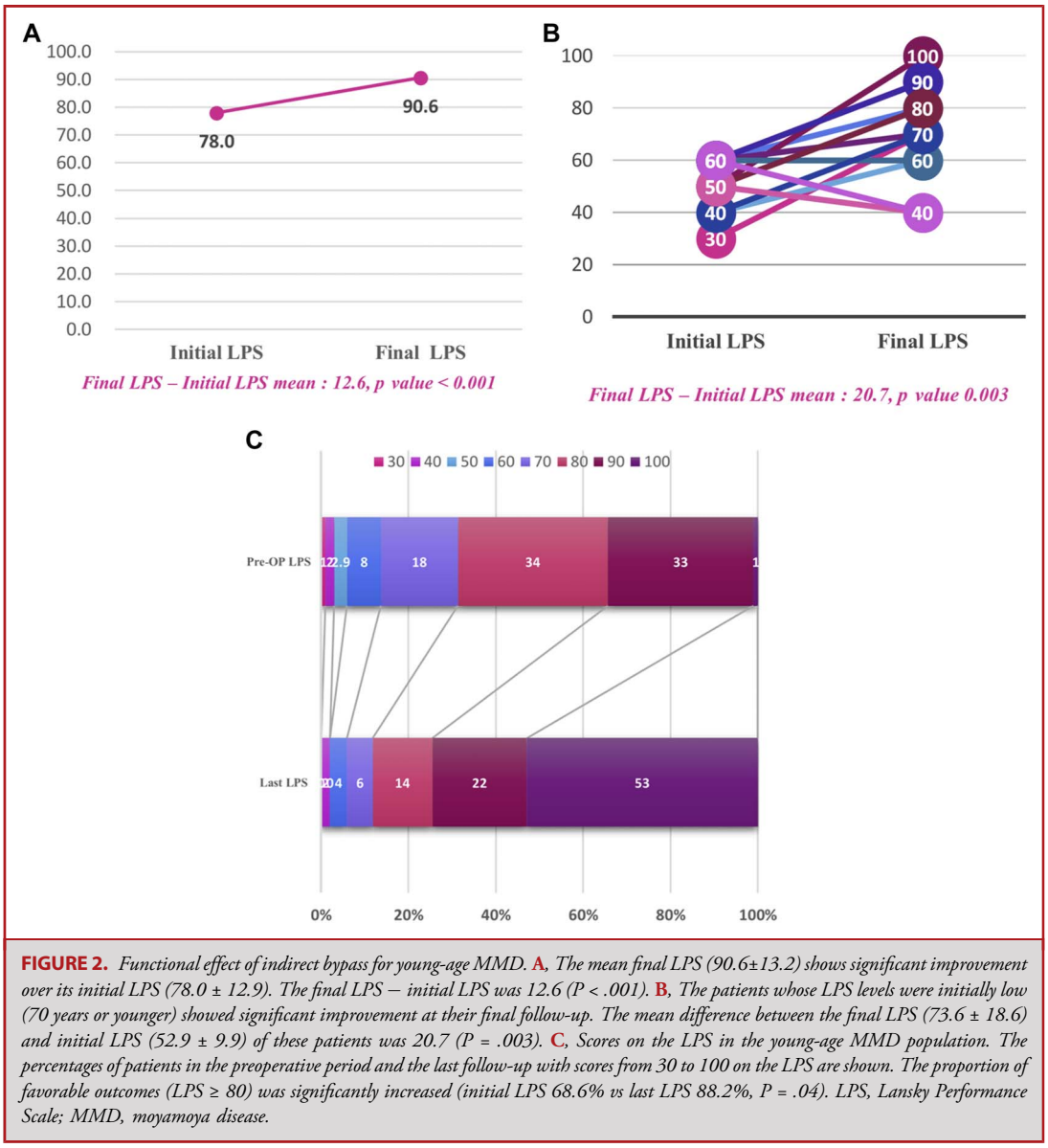

FIGURE 2. *Functional effect of indirect bypass for young-age MMD.* **A**, *The mean final LPS (90.6±13.2) shows significant improvement over its initial LPS (78.0 ± 12.9). The final LPS − initial LPS was 12.6 (P < .001).* **B**, *The patients whose LPS levels were initially low (70 years or younger) showed significant improvement at their final follow-up. The mean difference between the final LPS (73.6 ± 18.6) and initial LPS (52.9 ± 9.9) of these patients was 20.7 (P = .003).* **C**, *Scores on the LPS in the young-age MMD population. The percentages of patients in the preoperative period and the last follow-up with scores from 30 to 100 on the LPS are shown. The proportion of favorable outcomes (LPS ≥ 80) was significantly increased (initial LPS 68.6% vs last LPS 88.2%, P = .04). LPS, Lansky Performance Scale; MMD, moyamoya disease.*

13.2) showed significant improvement over its initial LPS (78.0 ± 12.9). The final LPS − initial LPS was 12.6 ($P < .001$, Figure 2A). Only 5% of patients showed functional decline. Even patients whose LPS was initially low (<80) showed significant improvement. The mean difference between the final LPS (73.6 ± 18.6) and initial LPS (52.9 ± 9.9) of these patients was 20.7 ($P = .003$, Figure 2B). There were also significant differences in good LPS between the initial and last follow-up (68.6% vs 88.2%, $P = .04$, Figure 2C).

Of the 102 patients followed up for more than 5 years, 44 (43%) had seizures at diagnosis. Of these patients, 38 (86%) were seizure-free and none are currently taking anticonvulsants.

Forty-two of 102 patients underwent follow-up K-WISC-IV, and 20 had borderline or higher intelligence (K-WISC-IV score 70 or higher). We confirmed the developmental and educational level of 42 patients who did not undergo K-WISC-IV through medical records and interviews. Of these, 31 said they had no problem completing the general education curriculum. That is, out of 84 patients, 33 (39%) showed mental retardation.

Factors Associated with Unfavorable Outcome (Table 5)

Risk factors such as younger than 3 years, initial presentation with symptomatic infarction, initial presentation with bilateral and multiple infarctions, postoperative complications, and the presence of PCA involvement were not associated with unfavorable outcome among younger than 4 years on univariate logistic regression analysis. The presence of subsequent preoperative infarction and initial LPS

TABLE 5. Univariate and Multivariate Logistic Regression Analyses of Risk Factors for Unfavorable Outcome (LPS < 80)

Characteristics	Parameters	OR	95% CI	*P* value
Univariate analysis				
Age at operation	<3	6.4	0.8-51.9	0.08
Initial presentation with symptomatic infarction	Present	6.1	0.7-49.5	0.09
Initial presentation with bilateral infarction	Present	3.4	0.9-12.5	0.07
Initial presentation with multiple infarction	Present	3.4	0.7-16.8	0.13
Subsequent preoperative infarction	Present	8.4	1.6-45.1	0.01
Initial LPS	<80	30.9	3.7-255.2	0.01
Postoperative complications	Present	2.6	0.7-9.3	0.15
PCA involvement	Present	0.9	0.2-3.8	0.91
Multivariate analysis				
Subsequent preoperative infarctions	Present	4.2	0.6-28.1	0.14
Initial LPS	<80	26.1	3.1-218.8	0.03

LPS, Lansky Play Performance Scale; OR, odds ratio; PCA, posterior cerebral artery.

was significant risk factors. However, multivariate logistic regression analysis revealed that initial LPS (<80) was most strongly associated with unfavorable outcomes (odds ratio [OR], 26.1; 95% CI, 3.1-218.8; $P = .03$). Bilateral and multiple cerebral infarctions at the time of onset were significantly associated with low initial LPS.

Longitudinal Analysis: Newly Developed Stroke Event and Stroke-free Survival Rate (Table 6)

The mean follow-up duration of 102 patients was 18.8 (5-27.3) years. During the follow-up period, a total of 8 infarctions occurred. Of these, 5 asymptomatic infarctions occurred at the site of the previous infarcted area. A total of 3 symptomatic infarctions occurred on the operated hemisphere. All new symptomatic infarctions occurred within 1 year after surgery (postoperative 3, 3, and 10 months each). New hemorrhage after surgery did not occur.

During the entire follow-up period, the overall newly developed symptomatic infarction rate was 2.9% (3/102). The annual symptomatic infarction rate was 0.16% per person-year. The 20-year event-free survival rate for infarction was 88% (Figure 3A). The 20-year event-free survival rate for symptomatic was 97% (Figure 3B).

TABLE 6. Longitudinal Analysis: Newly Developed Stroke Event and Stroke-Free Survival Rate (N = 102)

Parameters		No. of cases or value
Follow-up period		18.8 (5-27.3) y
New developed stroke event		
Infarction	Asymptomatic infarction	5
	Symptomatic infarction	3 (3, 3, 10 mo)
Hemorrhage		0
Annual symptomatic infarction rate		0.16%

DISCUSSION

Characteristics of Young-Age MMD

The 74% symptomatic infarction rate of young patients with MMD in this study is significantly higher than the 25% symptomatic infarction rate in pediatric patients with MMD.[6] Subsequent infarction frequently occurred despite a short waiting time of less than 2 weeks on average from the decision to perform surgery. The frequency of postoperative infarction is also higher in young patients with MMD than in older patients.[9,10]

The incidence of seizures is much higher in young patients with MMD. According to Mikami et al,[16] onset age 3 years or younger had higher rates of epilepsy than onset age 4 years or older (OR, 12.50; 95% CI, 1.00-20.00; $P = .008$). Of their cohort, 50% of the patients presented with seizure. They explain that the incidence of significant infarction is much higher in young-age MMD, which can naturally accompany seizures at a

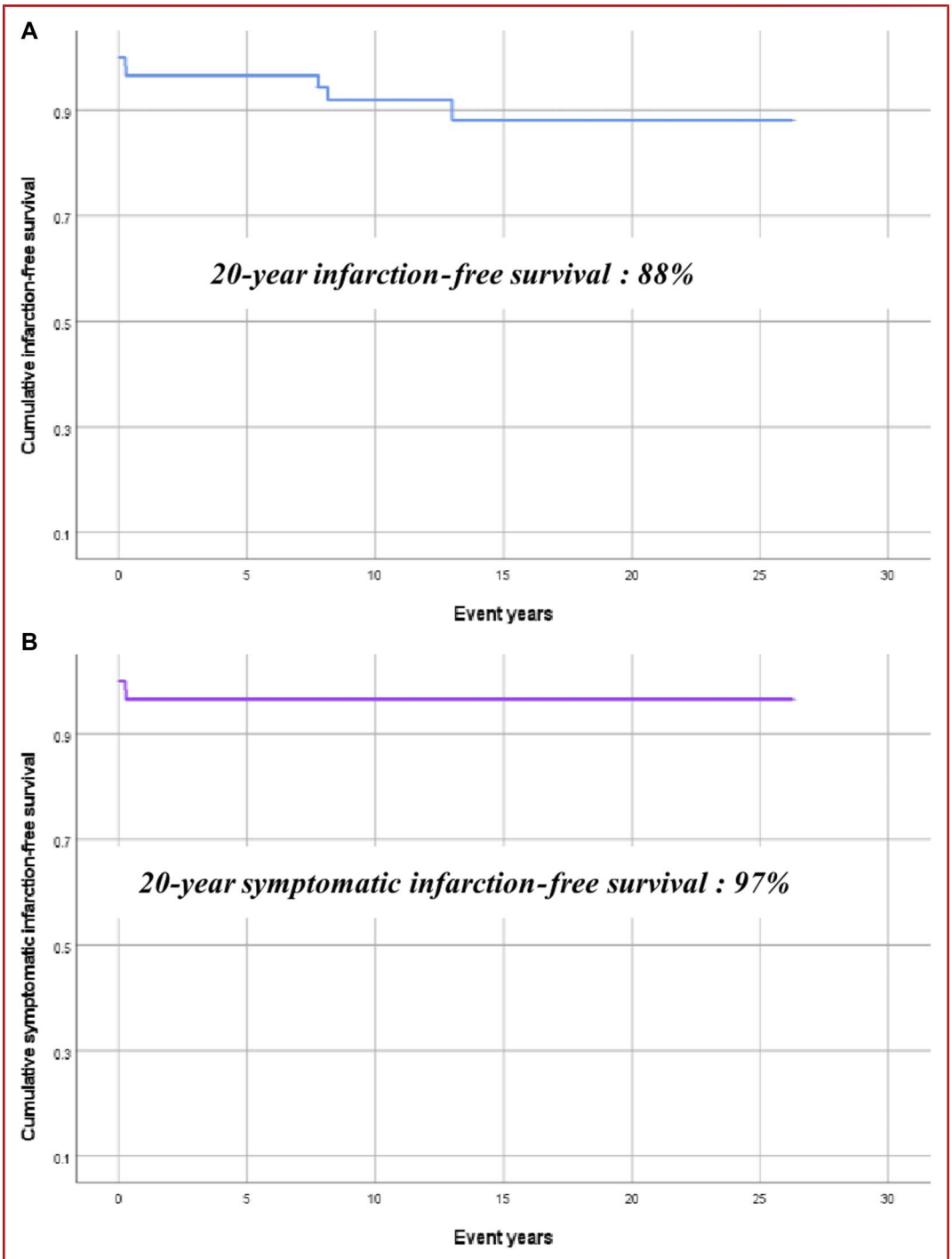

FIGURE 3. *Stroke prevention effect of indirect bypass for young age patients with moyamoya disease.* **A**, *Kaplan-Meier survival curve of infarction. The 20-year event-free survival rate for any infarction ipsilateral to the operated hemispheres was 88%.* **B**, *Kaplan-Meier survival curve of symptomatic infarction. The 20-year event-free survival rate for symptomatic infarction ipsilateral to the operated hemispheres was 97%.*

high rate. Seizures in MMD are presumed to be related to cerebral ischemia.[17,18] Young patients with MMD are more susceptible to cerebral ischemia due to the increased demand for cerebral blood flow (CBF) associated with brain development and insufficient supply due to poor collateral.[19,20] Owing to the characteristics of a young age (more infarction and vulnerability

to ischemia), seizure seem to be more frequent than other pediatric groups.

PCA stenotic progression was frequent (41%); even ICA stenosis was not severe in this age group. At the last follow-up, it was confirmed that 80 (59%) patients had PCA stenosis. PCA involvement in the pediatric MMD group was 15% to 20%.[14,21-23] PCA progression should be more carefully followed in young-age MMD.

Surgery for Young-Age MMD

Our previous study suggested the need for active surgical treatment in young patients with MMD.[9]

However, young age is a risk factor for surgical complications.[24] As postoperative infarction exacerbates the outcome, efforts should be made to minimize it. Our group has a management protocol.[9] Adequate preoperative hydration is the first step. During surgery, we maintain normal partial pressure of carbon dioxide ($PaCO_2$). Avoiding blood pressure drops and maintaining normal hemoglobin levels are also essential. Sufficient fluid supplementation is important. To minimize postoperative pain, a local nerve block is administered. Try to keep the patient from crying through pain and anxiety control.

Clinical Outcome of Young-Age MMD

In this study, 88% of patients had favorable clinical outcomes (final LPS 80 or higher). Furthermore, the final LPS level improved dramatically, suggesting that surgery is beneficial in this age group. LPS improved mainly because of the gain of motor function and the catch up of developmental delay. Since the developing brain has potent neuroplasticity, surgery would have augmented the neuroplastic potency by improving cerebral hemodynamics and preventing subsequent infarction.[25,26]

In young-age MMD, follow-up observations showed an overall recovery of 88%, showing better results than children's usual poststroke natural recovery process. Considering the natural course of MMD, the role of surgery is meaningful.

The seizure-free rate after surgery was excellent. After indirect bypass, most young patients with MMD with seizures benefited from increased cerebral perfusion in the MCA territory and a reduced risk of seizure recurrence.

Intellectual status after surgery seems disappointing. Our results showed that 39% of young patients with MMD had mental retardation postoperatively. Tagawa et al[27] reported that young age is associated with poor neuropsychological outcomes after surgery. Only 20% of patients (3 of 15) were normal postoperatively, and 6 of 7 symptomatic patients younger than 5 years had poor psychological results. Even in asymptomatic patients, cognitive function gradually decreases. Compared with the natural history, surgery might also be helpful in the cognitive aspect.[28-31]

In this study, considering the young age group, the sufficient period until adulthood was investigated. The stroke prevention effect was maintained up to 20 years after surgery. The 20-year event-free survival rate for symptomatic infarction in the operated hemisphere was 97%. Our previous study of a 629 pediatric MMD, 10-year event-free survival, or symptomatic infarction was 99%.[6]

All symptomatic cerebral infarctions in our patient group occurred within 1 year after surgery (3, 3, and 10 months, each). In other words, if enough collateral grows after indirect bypass, recurrent symptomatic infarction could be prevented in the long-term follow-up.

Factors Associated with Unfavorable Outcome

Bilateral and multiple cerebral infarctions at onset were significantly associated with low initial LPS. The initial poor functional status was the most critical factor on long-term poor outcome. It suggests that our strategy of preferentially operating the normal hemisphere before bilateral or multiple infarctions is a reasonable option.

There was no significant correlation between preoperative infarction and final clinical outcome. That is, it suggests that good functional recovery from infarct can be achieved by indirect bypass.

Limitations

MMD is a rare disease. The onset at a young age is even rarer. In addition to the small number of patients, there was a follow-up loss (4%), representing a limitation of a retrospective study that cannot rule out bias. Although this study is long-term follow-up results with an average of 18.8 years, it may be insufficient given the patient's overall life expectancy. More long-term follow-up data need to be accumulated to clarify the clinical course of young patients with MMD. Testing for Ring Finger Protein 213, known to be associated with young-age MMD and PCA progression,[32] was not performed in this study. If Ring Finger Protein 213 is tested in young-age MMD in the future, it might be helpful for the clinical management of these patients.

CONCLUSION

Young age in patients with MMD showed frequent symptomatic infarction, seizure at presentation, and PCA involvement. The incidence of subsequent infarction and the postoperative complication rate is also high. Even in this aggressive feature, indirect bypass surgery can improve long-term satisfactory clinical outcomes and prevent stroke effectively in young-age MMD.

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Disclosures

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COMMENTS

The authors present their experience spanning 32 years with moyamoya patients in the pediatric population. Specifically, they present 135 pediatric moyamoya patients under age 4. The long terms results are quite good in terms of stroke and cognitive outcome. They report an approximate 7% stroke risk per procedure with patients undergoing an average of 2.8 procedures. They study suffers from its retrospective nature and various interventions are lumped together, notwithstanding the results are excellent.

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The authors present a retrospective series of 135 pediatric moyamoya disease patients less than 4 years of age, that underwent an indirect bypass, with long term outcome in 102 children. They note a high overall complication rate, which may be unique to this population, but despite this they have excellent results in regards to their long term symptomatic stroke rate and their functional/cognitive outcomes.

The authors' data further reinforce that poor initial functional status is strongly associated with poor long-term outcome. This is consistent with our Stanford institutional moyamoya bypass data which shows higher perioperative complications in patients with radiographic perfusion signs of steal phenomenon after Diamox administration indicating severe disease, and suggests earlier surgery in moyamoya disease is warranted if possible.

Another factor to consider, which is very institutionally dependent, is the significant variety of surgeries that were performed. Though the authors point out the variance is due to degree of posterior circulation or ACA involvement, there still remains a significant number of patients which had an additional third or potentially a fourth operation. As the authors' overall complication rate is 15% per operation, it is unclear if some of these risks could be mitigated by performing a less-is-more approach, without the need of an additional bifrontal craniotomy or occipital EDAS in addition to the primary STA EDAS, that would not adversely affect the excellent 20-year infarction free survival.

The authors should be congratulated for analyzing the outcomes of revascularization in this interesting moyamoya subpopulation. Further research in this young pediatric moyamoya group should be undertaken.

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