Moyamoya Disease and Pregnancy: Case Report and Review of the Literature

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OBJECTIVE AND IMPORTANCE: Many female patients with moyamoya disease are of childbearing years, including those who were diagnosed before entering their childbearing years. However, there have been no extensive reviews of the management of pregnancy and delivery in association with moyamoya disease. The purpose of this report is to describe the case of a patient with moyamoya disease complicated by pregnancy and to review the literature on other such cases.

CLINICAL PRESENTATION AND INTERVENTION: We report a 23-year-old primipara with moyamoya disease who delivered uneventfully by cesarean section under spinal anesthesia at 38 weeks of gestation. In the literature, 30 cases were reported of patients who had been diagnosed with moyamoya disease before pregnancy and delivery, and 23 patients who were symptomatic and were diagnosed for the first time with moyamoya disease in association with pregnancy.

CONCLUSION: There is no evidence that pregnancy increases the risk of cerebrovascular accident or that bypass surgery decreases its risk. Poor prognosis of the patient or the newborn is mostly caused by cerebral hemorrhage and not by cerebral ischemia. It is important to control blood pressure and especially to avoid toxemia during pregnancy. Either cesarean section or vaginal delivery can be accomplished safely. Any anesthetic method can be used, provided special attention is given to avoiding hypocapnia, hypotension, and hypertension. Oral contraceptives should be avoided. (Neurosurgery 43:360–369, 1998)

Key words: Anesthesia, Cerebrovascular accident, Cesarean section, Delivery, Moyamoya disease, Pregnancy

Moyamoya disease as a clinical entity has been known for more than 30 years and is predominant in the Japanese population. Its cause, however, is still unknown. In general, pediatric patients with the disease present with ischemic symptoms, including seizure and involuntary movement, whereas adult patients present with hemorrhagic symptoms. Extracranial-intracranial (EC-IC) bypass surgery has been used to treat patients with ischemic symptoms (25, 33). For patients with hemorrhagic symptoms, however, the efficacy of such bypass surgery has not yet been established.

The incidence of moyamoya disease is higher in female than in male patients (15). Thus, among patients with moyamoya disease, women in their childbearing years are not uncommon, including those diagnosed before reaching childbearing age. Pregnant women with moyamoya disease may be divided into two categories in terms of the timing of clinical manifestation: those for whom diagnosis was made before the patient became pregnant (the patients known to have moyamoya disease) and those for whom diagnosis was made for the first time while the patient was pregnant or during puerperium (the patients newly diagnosed with moyamoya disease). In addition to participating in the clinical management of the patients in these two categories, neurosurgeons must be prepared to provide sufficient information regarding the risks associated with pregnancy to nonpregnant patients who are known to have moyamoya disease and who want to have a baby. Although there are case reports of pregnant patients newly diagnosed with moyamoya disease who present, in most cases, with intracranial hemorrhage and of delivery by patients known to have moyamoya disease, there have been no extensive reviews of the management of pregnancy and delivery in association with moyamoya disease. Furthermore, no guidelines on pregnancy management and contraception for patients known to have moyamoya disease have been issued. In this report, we add the case of pregnancy of a patient known to have moyamoya disease and review the literature on moyamoya disease in association with pregnancy.

CASE REPORT

This 23-year-old woman experienced a transient ischemic attack with right hemiparesis, including the face, when she was 6 years old. At the age of 7 years, a diagnosis of moyamoya disease was made using cerebral angiography because of repeated transient left hemiparesis. The patient's history and the histories of her family members were not remarkable. Duraencephalosynangiosis was performed bilaterally. The patient was given aspirin for several years, which was eventually discontinued. The patient had no history of cerebral ischemia, seizure, or cerebral hemorrhage thereafter. She was married at

the age of 22 years and came to our gynecological department when she was primigravid. Her menstrual history was not remarkable. Magnetic resonance (MR) angiography at the age of 21 years had revealed occlusion of the distal ends of the bilateral internal carotid arteries and collaterals from the external carotid system (*Fig. 1*). During pregnancy, there were no cerebrovascular accidents or toxemia, and development of the fetus and placenta were both normal.

The patient was admitted for management of delivery at 38 weeks of gestation. The results of general and neurological examinations were normal, with a blood pressure of 132/86 mm Hg. Cesarean section was performed on Day 4 with the patient under spinal anesthesia using 10 mg of tetracaine. To prevent hypotension, 10 mg of ephedrine was administered intravenously twice during the delivery. From the time of rupture of the amnion, the newborn was delivered in 1 minute and the placenta was delivered in 2 minutes. The male newborn had a birth weight of 2955 g and Apgar scores of 6 and 9. During the procedure, there was no observation of hypertension or hyperventilation. The operation time was 38 minutes, and that of anesthesia was 1 hour. During delivery and postpartum, no neurological deficits were noted. The patient and her baby were discharged without any problem on Day 8.

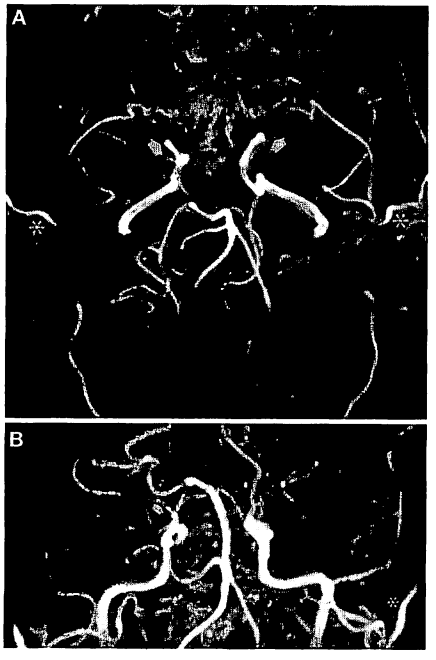

FIGURE 1. Axial view (*A*) and anteroposterior view (*B*) MR angiograms. The distal portion of the internal carotid arteries (*arrows*) is not visualized bilaterally. Neither are proximal portions of the bilateral middle cerebral arteries demonstrated. Enlarged superficial temporal arteries (*asterisks*) and middle meningeal arteries are observed bilaterally.

SUMMARY OF THE REPORTED CASES

Because the clinical manifestation and management of patients known to have moyamoya disease are completely different from those of patients newly diagnosed with moyamoya disease, we describe these two groups separately.

Pregnancy, delivery, puerperium, and abortion among patients known to have moyamoya disease (*Table 1*)

Including our patient (Patient 30), 30 patients (31 deliveries) fall into this category. Patient age ranged from 20 to 36 years, with an average of 26.7 years. Although Patients 18, 23, and 29 were secundipara, the rest of the patients were primipara except for Patient 21, who underwent two vaginal deliveries. Diagnosis of moyamoya disease was made at ages ranging from 3 to 32 years, with an average of 17.4 years. The initial symptom was cerebral ischemia, including seizure and involuntary movement, in 17 patients. This occurred at ages ranging from 3 to 28 years, with an average of 13.9 years. Cerebral hemorrhage was the initial symptom in 10 patients, occurring at ages ranging from 16 to 32 years, with an average of 22.9 years. Surgical treatment was performed on 12 patients, whereas no surgical treatment was performed on 18 patients. Surgical treatment was performed on six patients with hemorrhagic onset and on three patients with ischemic onset. In the case of Patient 15, who had ischemia at age 12 years and hemorrhage at age 16 years, unilateral superficial temporal artery to middle cerebral artery anastomosis was performed. Unilateral superior cervical ganglionectomy was performed on Patient 5. In 11 patients, EC-IC bypass surgery, either superficial temporal artery to middle cerebral artery anastomosis or duraencephalosynangiosis, was performed, but this surgery was not performed bilaterally in all cases. Cesarean section was performed in 25 patients, whereas vaginal delivery was accomplished in 5 patients (six deliveries). The anesthetic methods were general in 6 patients, epidural in 12, spinal in 10, and pudendal in 1.

Poor outcome among the patients known to have moyamoya disease was observed only in Patient 16, a 23-year-old primipara who, at the age of 10, had been diagnosed with moyamoya disease, presenting with ischemic symptoms. This patient, who had not undergone EC-IC bypass, developed bilateral ventricular hemorrhage at 30 weeks of gestation. After cesarean section, bilateral ventricular drainage was performed. The baby developed normally, but the patient entered a state of akinetic mutism.

There were two cases of uneventful delivery in which the newborns had congenital anomalies. Patient 1 was a 24-year-old primipara who had been prescribed cyclandelate and pyridinol carbamate. These were discontinued when her pregnancy became known. The baby was born with a cleft palate. Patient 2 was a 34-year-old primipara, who had been prescribed phenytoin and phenobarbital during the period from 23 to 34 weeks of gestation. The baby was born with osteogenesis imperfecta of the cranium and the femur, and the bilateral humerus fractured pathologically at 2 weeks postpartum.

In four patients (Patients 2, 11, 15, and 17), transient ischemic attack or reversible ischemic neurological deficits of hemiparesis or monoparesis occurred during pregnancy or postpartum, but all of these patients achieved good outcomes. In Patient 10, generalized seizure occurred on six occasions between 18 to

TABLE 1. Pregnancy, Delivery, Puerperium, and Abortion among Patients Known to Have Moyamoya Disease[a]

Patient no.	Series (Ref. No.)	Age (yr)	Para	Diagnosis (age in yr)	Initial Symptoms	Surgical Treatment	Toxemia	Gestation (wk)	Delivery	Anesthesia
1	Matsuoka et al., 1976 (32)	24	0	19	I	No		40	CS	General
2	Matsuoka et al., 1976 (32)	34	0	26	I	No		40	CS	General
3	Bingham et al., 1980 (6)	25	0	21	I	No		?	VD	Pudendal
4	Sasaki et al., 1984 (46)	30	0	28	I	No		38	CS	Spinal
5	Sera et al., 1985 (47)	31	0	16	H	Ganglionectomy/L	Yes	39	CS	Spinal
6	Urushikawa, 1986 (60)	28	0	23	H	No		33	VD	No
7	Miyakawa et al., 1986 (34)	29	0	19	H	No	Yes	38	CS	Epidural
8	Fukada et al., 1988 (13)	30	0	24	H	No		38	CS	Epidural
9	Tadakuma et al., 1989 (51)	27	0	25	H	No		38	CS	Epidural
10	Nakago et al., 1990 (39)	24	0	10	I	No		36	CS	General
11	Saruki et al., 1990 (45)	28	0	10	I	No		38	CS	General
12	Saruki et al., 1990 (45)	21	0	3	I	No		38	CS	Epidural
13	Fayle and Armatage, 1992 (9)	24	0	16	Hemiparesis?	No		40	VD	Epidural
14	Murase et al., 1992 (37)	29	0	22	I	Anastomosis	Yes	38	CS	Spinal
15	Kase et al., 1993 (26)	26	0	12	I and H	STA-MCA/R		37	CS	General
16	Shimamoto et al., 1994 (49)	23	0	10	I	No		30	CS	General?
17	Sharma et al., 1994 (48)	26	0	21	I	No		38	CS	Epidural
18	Venkatesh and Taggart, 1994 (62)	?	1	?	?	?		?	CS	General
19	Hara et al., 1994 (16)	20	0	10	I	No		38	CS	Epidural
20	Tomoda and Ogita, 1995 (58)	22	0	19	Seizure	Yes		39	CS	Epidural
21	Tomoda and Ogita, 1995 (58)	20	0	17	H	Yes		39	VD	Epidural
		22	1	17	H	Yes		39	VD	Epidural
22	Tomoda and Ogita, 1995 (58)	30	0	4	Hemiparesis/aphasia	Yes	Yes	40	VD	Epidural
23	Tomoda and Ogita, 1995 (58)	29	1	28	H	Yes		38	CS	Spinal
24	Terauchi et al., 1995 (57)	35	0	25	H	EDAS/B		38	CS	Spinal
25	Terauchi et al., 1995 (57)	31	0	20	H	EDAS/R		38	CS	Spinal
26	Terauchi et al., 1995 (57)	22	0	4	I	EDAS/B		38	CS	Spinal
27	Terauchi et al., 1995 (57)	28	0	9	I	No		38	CS	Spinal
28	Miyake et al., 1996 (35)	25	0	7	I	No		38	CS	Spinal
29	Kee and Gomersall, 1996 (27)	36	1	32	H	No		37	CS	Epidural
30	Present study	23	0	6	I	EDAS/B		38	CS	Spinal

[a] **I, ischemia; H, hemorrhage; ?, unknown; CS, Cesarean section; VD, vaginal delivery; L, left side; R, right side; B, bilateral; STA-MCA, superficial temporal artery to middle cerebral artery anastomosis; EDAS, encephaloduromyosynangiosis; TIA, transient ischemic atta**

31 weeks of gestation. The patient experienced additional seizures immediately postpartum, after 24 hours, and 3 days later. Both the patient and the baby were finally discharged in good condition.

For Patients 10 and 15, cesarean section was performed with the patients under general anesthesia. Both newborns were sleeping babies but were discharged without any sequels. For Patient 6, spontaneous vaginal delivery without anesthesia was accomplished. The baby experienced seizure 3 hours immediately postpartum because of ventricular hemorrhage, but the outcome was good.

Patient 2 had spontaneous abortion at the gestation period of 4 months because of intrauterine fetal death. Patient 13 underwent therapeutic abortion at 13 weeks of gestation without any sequel.

Maternal Events	Fetal Events	Maternal Outcome	Fetal Outcome	Remarks
No	Cleft palate	Good	Good	Medication
Hemiparesis and abnormal sensation at 23 wk	Osteogenesis imparfecta	Good	?	Previous spontaneous abortion, medication
No	No	Good	Good	Epidural anesthesia failed, forceps delivery
Occasional migraine during pregnancy	No	Good	Good	Unilateral moyamoya disease
No	No	Good	Good	Breech presentation
No	Ventricular hemorrhage	Good	Good	Newborn had seizure 3 h postpartum
No	No	Good	Good	Subsequent bleeding at 7 mo, EDAS at 9 mo postpartum
No	No	Good	Good	
No	No	Good	Good	
Seizure at 18–31 wk, immediately postpartum, at 24 h, at Day 3	Sleeping baby	Good	Good	Seizure caused by hypovolemia, hypocapnia
TIA (monoparesis)	No	Good	Good	Left stellate ganglion block
Vomiting and headache at 30 wk	No	Good	Good	Headache and right hand weakness at 1 wk
No	No	Good	Good	Therapeutic abortion at 13 wk at 23 yr
No	No	Good	Good	
TIA on Days 2 and 4	Sleeping baby	Good	Good	Ischemia at 12 yr, putaminal hemorrhage at 16 yr
Ventricular hemorrhage at 30 wk	No	Akinetic mutism	Good	Ventricular drainage and emergency CS
Increased hemiparesis, dysarthria at 31 wk	No	Good	Good	
No	No	Good	Good	Previous CS under general anesthesia
No	No	Good	Good	
No	No	Good	Good	Anastomosis at 20 yr
No	No	Good	Good	
No	No	Good	Good	Forceps delivery, previous delivery 2 yr previously
No	No	Good	Good	Forceps delivery
No	No	Good	Good	Previous CS
Abnormal sensation in early pregnancy	No	Good	Good	
No	No	Good	Good	
No	No	Good	Good	
No	No	Good	Good	
No	No	Good	Good	Hypertension and hyperventilation during CS
No	No	Good	Good	
No	No	Good	Good	1 normal delivery, 13 yr ago

Pregnancy, delivery, puerperium, and abortion among patients newly diagnosed with moyamoya disease (*Table 2*)

In the literature review, 23 patients fall within this category. Patient age ranged from 23 to 35 years, with an average of 28.3 years. Four patients (Patients 37, 40, 44, and 50) were secundipara and 12 patients were primipara. Cerebral hemorrhage occurred in 16 patients, and cerebral ischemia, including seizure and involuntary movement, occurred in 3 patients (Patients 45, 46, and 48). Cerebral hemorrhage occurred at gestation periods ranging from 15 to 37 weeks, with an average of 28.1 weeks. Patient 53, who underwent spontaneous vaginal delivery at 40 weeks of gestation, had cerebral hemor-

TABLE 2. Pregnancy, Delivery, Puerperium, and Abortion among Patients Newly Diagnosed with Moyamoya Disease[a]

Patient No.	Series (Ref. No)	Age (yr)	Para	Ictus	Symptoms	Surgical Treatment	Toxemia
31	Takagi et al., 1977 (52)	35	0	During delivery	H/I?	No	
32	Fujita et al., 1978 (12)	30	1?	7 mo	H	Hematoma removal	
33	Karasawa et al., 1980 (24)	27	?	9 mo	H	No	
34	Hashimoto et al., 1985 (18)	28	?	34 wk	H	?	
35	Hashimoto et al., 1985 (18)	27	?	24 wk	H	?	
36	Fukuo et al., 1987 (14)	30	0	20 wk	H	No	
37	Enomoto and Goto, 1986 (8)	32	1	32 wk	H	Hematoma removal/EDAS	
38	Teramoto et al., 1987 (56)	28	0	37 wk	H	Hematoma removal	Yes
39	Hashimoto et al., 1988 (17)	24	0	6 mo	H	STA-MCA/L	
40	Iwasato et al., 1989 (22)	28	1	33 wk	H	Hematoma removal/stereotaxy	Yes
41	Ohtsuka and Mori, 1989 (42)	28	?	4 mo	Involuntary movement	No	
42	Negi et al., 1990 (41)	28	?	24 wk	H	Ventricular drainage	
43	Negi et al., 1990 (41)	32	?	25 wk	H	No	
44	Miyauchi et al., 1991 (36)	30	1	36 wk	H	Ventricular drainage	Yes
45	Kikukawa et al., 1991 (29)	24	0	During delivery	I (seizure)	STA-MCA, EMS/B	
46	Yajima and Mita, 1992 (64)	23	0	23 wk	I	No	
47	Nanato et al., 1992 (40)	24	0	During delivery	Seizure	No	
48	Ushimura et al., 1993 (61)	31	0	4 mo	Blindness	No	
49	Amin-Hanjani et al., 1993 (2)	25	0	?	H?	No	
50	Henmi et al., 1993 (19)	31	1	15 wk	H	Ventricular drainage	
51	Takeda et al., 1994 (54)	25	0	27 wk	H	Ventricular drainage	
52	Umeki et al., 1995 (59)	31	0	27 wk	H	Ventricular drainage	
53	Tanioka et al., 1995 (55)	29	0	4.5 h postpartum	H	Hematoma removal	

[a] **H, hemorrhage; I, ischemia; ?, unknown; CS, Cesarean section; VD, vaginal delivery; EDAS, encephaloduromyosynangiosis; STA-MCA, superficial temporal artery to middle cerebral artery anastomosis; L, left side; R, right side; B, bilateral; EMS, encephalomyosynangiosis; TIA, transient ischemic attack.**

rhage 4.5 hours immediately postpartum. Ischemic episodes occurred at periods ranging from 4 months to 40 weeks of gestation. Patients 45 and 47 developed generalized seizure during delivery. Patient 31 experienced cerebrovascular accident during delivery. Cesarean section was performed in 14 patients, whereas vaginal delivery was accomplished in 4 patients. Cesarean section was performed at stages ranging from 27 to 40 weeks of gestation, with an average of 34.6 weeks. Vaginal delivery occurred between at 35 and 40 weeks of gestation. Delivery was attempted immediately after cerebrovascular accidents in the cases of Patients 38, 44, and 51, but delivery was delayed in 10 patients. Patients 32 and 46 underwent therapeutic abortion. This induced cerebral ischemia in Patient 46, with psychiatric manifestation. The anesthetic methods were general in seven patients, epidural in three, and spinal in two. The outcomes of the mothers were 3 deaths, 6 poor recoveries, 2 akinetic mutisms, and 10 good recoveries. Patient 43 died as a result of repeated hemorrhage occurring at 25 and 30 weeks of gestation. The outcomes of the newborns were 2 deaths (another death was the result of therapeutic abortion), 13 good recoveries, and 1 hemiparesis. With one exception, all of the deaths of the mothers and the newborns and the maternal poor prognosis were the result of cerebral hemorrhage. The single exception, Patient 46, manifested psychiatric symptoms caused by cerebral infarction of the frontal lobe, which was caused by therapeutic abortion.

In patients with cerebral hemorrhage, removal of the hematoma through craniotomy was performed in four patients (Patients 32, 37, 38, and 53), whereas ventricular drainage was performed in five patients. In Patient 40, stereotactic removal of the hematoma was attempted but was unsuccessful. In Patients 37, 39, and 45, EC-IC bypass surgery was performed. No surgical treatment was performed in nine patients.

Patient 44 had previously had a spontaneous abortion. In Patients 38, 40, and 44, pregnancy was complicated by toxemia, which might have been related to cerebral hemorrhage.

DISCUSSION

Pregnancy and cerebrovascular accidents

During pregnancy, circulating blood volume increases acutely, by 30 to 60% from the first to second trimester, over that in the nonpregnant state. This increase in blood volume occurs not in the brain or liver but in the uterus, kidneys, and limbs (28). Intracranial hemorrhage during pregnancy is caused in most cases by cerebral aneurysms, cerebral

Gestation	Delivery	Anesthesia	Maternal Outcome	Newborn Outcome	Remarks
40 wk	CS	Spinal	Poor	Good?	Consciousness disturbance and epilepsy during delivery
7 mo	Abortion	General	Akinetic mutism	Death	Intrauterine fetal death
35 wk	VD	Epidural	Impaired intelligence	Good	Delivery with vacuum extractor
?	?	?	?	?	
?	?	?	?	?	
37 wk	CS	Epidural	Good	Good	Vaginal delivery attempted
38 wk	VD	?	Good	Good	First delivery 3 yr previously
37 wk	CS	General	Akinetic mutism	Good	Death as a result of subsequent bleeding 11 mo later
39 wk	VD	Epidural	Good	Good	Vacuum delivery, akin moyamoya disease
34 wk	CS	Spinal	Aphasia/hemiparesis	Good	Unsuccessful stereotactic hematoma removal
?	?	?	Good	Good	Involuntary movement recurred 2 mo postpartum
30 wk	CS	?	Good	Death	Intrauterine fetal death at 30 wk, STA-MCA later
30 wk	CS	General?	Death	Hemiparesis	Subsequent bleeding at 30 wk
36 wk	CS	General	Poor	Good	Diagnosed on Day 33, EDAS/R on Day 57
40 wk	CS	?	Good	Good	Fetal distress prompted CS
23 wk	Abortion	Intravenous?	Poor	(Death)	Therapeutic abortion induced infarction
39 wk	CS	General	Good	Good	Seizure during labor
34 wk	CS	General	Good	Good	
?	CS	General	Good	?	Seizure probably caused by hemorrhage
38 wk	CS	General	Good	Good	Quadrantanopsia
27 wk	CS	?	Death	?	Apgar 5/8
28 wk	CS	?	Death	?	Apgar 3
40 wk	VD	None	Poor	Good	Spontaneous delivery

arteriovenous malformations, or toxemia (1, 20, 43). Indication of neurosurgical intervention for intracranial hemorrhage during pregnancy is generally the same as in the nonpregnant women (1, 12, 43). For ruptured cerebral aneurysms, clipping is performed, principally because subsequent bleeding may be fatal. For ruptured arteriovenous malformations, surgery or conservative treatment is selected, although there remains some risk of subsequent bleeding with conservative treatment. The risk of cerebral ischemia during pregnancy increases by factors ranging from 3 to 13 over that in a nonpregnant state (23, 63). In general, arterial occlusion occurs during the second and third trimesters of pregnancy or the 1st week postpartum, whereas cerebral venous sinus occlusion occurs between 1 and 4 weeks postpartum (63). In the treatment of cerebral ischemic disease, warfarin should be avoided because of its teratogenic effects and fetal wastage. Instead, heparin or aspirin, which have no teratogenic effects, may be used (63).

Pregnancy in patients with moyamoya disease

During pregnancy of patients with moyamoya disease, it is important to prevent cerebral ischemia and cerebral hemorrhage. Furthermore, not only the life of the mother but fetal lives as well should be protected. The initial symptoms of cerebral hemorrhage are headache, consciousness disturbance, hemiparesis, generalized seizure, and hypertension. These should be differentiated from other cerebrovascular diseases or toxemia, including eclampsia (52). Some patients are first misdiagnosed as having eclampsia and are later proved to have moyamoya disease (36, 52). Computed tomography is necessary for diagnosis when cerebrovascular disease is suspected. Cerebral angiography is required when cerebral aneurysms, cerebral arteriovenous malformations, or other cerebrovascular diseases are suspected (1, 20). MR angiography is noninvasive and of value for the diagnosis of moyamoya disease, even early in pregnancy.

Including our case reported herein, there are 53 patients with moyamoya disease associated with pregnancy reported in the literature. However, some patients (6, 17, 46) do not satisfy the diagnostic criteria of moyamoya disease established by the Research Committee on Spontaneous Occlusion of the Circle of Willis (moyamoya disease) of the Ministry of Health and Welfare, Japan. These diagnostic criteria are as follows: 1) bilateral steno-occlusive changes at the distal portions of the internal carotid arteries and the proximal portions of the anterior and/or middle cerebral arteries, and 2) abnormal fine networks of

vessels (known as moyamoya vessels) at the base of the brain.

Hemorrhage in moyamoya disease is supposed to occur from the following: 1) saccular aneurysm in the circle of Willis; 2) small peripheral pseudoaneurysm, usually arising from the perforating arteries or choroidal arteries; or 3) rupture of the fragile moyamoya vessels in the basal ganglia (3, 24, 38). There are no reports of saccular aneurysms or pseudoaneurysms in association with pregnant moyamoya patients. If hemorrhage is caused by a saccular aneurysm, clipping of the aneurysm should be performed. If hemorrhage is caused by a pseudoaneurysm, it is usually treated conservatively. Most intracerebral hemorrhage occurring in association with moyamoya disease is intraventricular hemorrhage or hemorrhage at the basal ganglia or thalamus. Incidence of initial hemorrhage or subsequent bleeding is higher in female than in male patients (3, 44).

Although there are only a few reports of intracranial hemorrhage in association with pregnancy, there is 1 patient among the known patients with moyamoya disease and 14 patients among those newly diagnosed with moyamoya disease. Cerebral hemorrhage occurs during the second or third trimester of pregnancy, similar to the occurrence of aneurysmal subarachnoid hemorrhage during pregnancy (1, 43) (*Fig. 2*). Cerebral ischemia occurred during pregnancy in four of the patients known to have moyamoya disease and in three of the patients newly diagnosed with moyamoya disease. There is no evidence that pregnancy in patients with moyamoya disease increases the risk of cerebral hemorrhage or cerebral ischemia. However, the increase in circulating blood volume, hypercoagulability, and the complication of toxemia may cause deterioration of the clinical symptoms of moyamoya disease. Thus, patients known to have moyamoya disease should be made aware of the potential risks of pregnancy. Among the reported patients known to have moyamoya disease, poor prognosis was noted in only one patient with ventricular hemorrhage (49). The remaining patients had a good prognosis. The prognosis of reported nonpregnant patients with moyamoya disease (both male and female patients) with cerebral hemorrhage was good in 60% of the cases, whereas the mortality rate ranged from 14 to 22% (44). Predicting and preventing cerebral hemorrhage, some authors counsel patients who are known to have moyamoya disease to avoid pregnancy (35, 36, 46, 56).

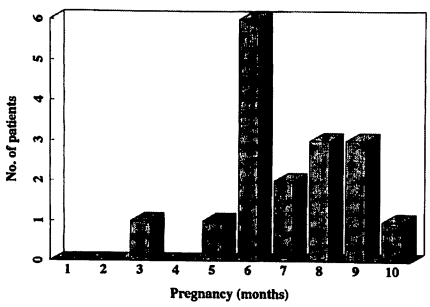

FIGURE 2. Cerebral hemorrhage among patients with moyamoya disease during pregnancy. Cerebral hemorrhage occurs mainly in the second and third trimesters.

A poor prognosis in cases of pregnancy in association with moyamoya disease is, in most cases, caused by cerebral hemorrhage (57) and not by cerebral ischemia. Although EC-IC bypass surgery is recommended before pregnancy (39) or even after intracerebral hemorrhage during pregnancy (57), the efficacy of EC-IC bypass surgery for the prevention of cerebral hemorrhage is not yet established. Furthermore, there are no cases of cerebral ischemia in the patients known to have moyamoya disease resulting in poor prognosis. Thus, surgical indication of EC-IC bypass should be critically assessed. One patient developed transient ischemic attack 2 days postpartum, even with treatment by EC-IC bypass surgery (53). It is important to control blood pressure, especially to prevent toxemia that could result in cerebral hemorrhage. Excessive exercise during pregnancy, including gymnastics and swimming, is contraindicated (54).

Contraception, therapeutic abortion and repeated pregnancies among patients with moyamoya disease

Oral contraceptive drugs caused deterioration of the clinical symptoms of patients with moyamoya disease (7, 21, 31, 50). The drugs themselves, however, do not always increase the risks of cerebral ischemia (23). For patients known to have moyamoya disease, condoms or intrauterine devices are recommended as contraceptives and oral contraceptive drugs are to be avoided (9, 46). Therapeutic abortion can be performed (9, 22), but this is accompanied by some risk of cerebrovascular accidents (64). Fujita et al. (12) proposed that the indications for therapeutic abortion for patients with intracranial hemorrhage are as follows: 1) the general condition and neurological status of the patient are serious; and 2) surgical treatment is difficult, and subsequent bleeding is likely. There is only one patient known to have moyamoya disease (58) who has had repeated (two) deliveries. It is unknown whether repeated pregnancy increases the risk of cerebrovascular accidents.

Methods of delivery among patients with moyamoya disease

Intentional delivery before the estimated date of confinement is recommended to reduce the risks to the mother when the fetus becomes mature (32, 46). Cesarean section is recommended to prevent hypertension caused by labor in the second stage of vaginal delivery, and cerebral ischemia caused by hyperventilation (32, 46, 57). However, cesarean section may be dangerous because of rapid circulatory changes at the laparotomy and at the delivery. Vaginal delivery with forceps under epidural anesthesia can be used to reduce the stress to the cardiovascular system (9, 58). Because intracranial hemorrhage at delivery rarely occurs in patients with cerebral aneurysms or cerebral arteriovenous malformations, vaginal delivery is recommended (1). Vaginal delivery under either epidural or spinal anesthesia is possible when the birth canal is soft and delivery proceeds rapidly (46). Among the reported patients, none of the patients known to have moyamoya disease developed cerebral hemorrhage or cerebral ischemia during delivery whereas one of the patients newly diagnosed with moyamoya disease developed cerebrovascular accident and another developed generalized seizure during delivery as the initial symptoms. Thus, we think that it is unnecessary to insist on cesarean section for patients known to have moyamoya disease. Because uterine contractile drugs dur-

ing puerperium may occasionally cause hypertension, they should be used with great caution (45, 46). The case of a patient who bled immediately postpartum (55) suggests the importance of controlling blood pressure and pain, even postpartum.

Methods of anesthesia for patients with moyamoya disease

During EC-IC bypass surgery in patients with moyamoya disease, hypocapnia may cause cerebral ischemia, and normal or slightly raised arterial carbon dioxide pressure is recommended (5, 30). Anesthesia used in delivery in reported cases of patients with moyamoya disease has been either general (11, 16, 26, 53, 62), epidural (16, 27, 48), or spinal (57). The usefulness of stellate ganglion block (45) is questionable. With any method of anesthesia, it is important to maintain cerebral blood flow and stable blood pressure while avoiding hyperventilation, hypotension, or hypertension. General anesthesia may be dangerous because of hypertension at intubation, aspiration of stomach contents, or neonatal depression (27, 48, 58). Two newborns were sleeping babies (26, 39, 53) but without any sequels. Spinal or epidural anesthesia allows continuous monitoring of neurological status during the operation, but hypotension must be carefully avoided (48). Light sedation is required to eliminate the patient's anxiety (16). One patient developed hypertension and hyperventilation because of anxiety during cesarean section (35). In the first stage of labor, hyperventilation caused by pain may cause hypocapnia, but epidural anesthesia may maintain arterial carbon dioxide pressure in the normal range (10) and is even useful for pain relief postpartum (35, 45, 48, 58). Monitoring the arterial pressure and arterial carbon dioxide pressure is indispensable, and maintaining body temperature is also important. Judging from the outcomes of the reported cases, it is not the method of anesthesia but the safe and careful anesthetic procedures that contribute most to the successful management of delivery among patients with moyamoya disease.

Newborns

Congenital anomaly (cleft palate, osteogenesis imperfecta, and maldevelopment of the femur) was observed in two cases involving mothers who had been prescribed several drugs during pregnancy (32). It is not known whether these anomalies were associated with moyamoya disease or with the drugs. Because there are reports of familial occurrence of moyamoya disease (15, 50), a noninvasive diagnostic examination, such as MR imaging and/or MR angiography (4), is recommended for the offspring of a patient with moyamoya disease when the child becomes older.

CONCLUSION

We think that cooperation among neurosurgeons, gynecologists, and anesthesiologists is important for the successful management of pregnancy and delivery for patients with moyamoya disease.

Received, December 30, 1997.
Accepted, April 1, 1998.
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COMMENTS

The authors present the case of a young woman who had a diagnosis of moyamoya disease confirmed angiographically at age 7 years. At age 23 years, she presented at 38 weeks gestation and subsequently underwent caesarian section. Her course was uneventful. In reviewing the records and case reports of 30 other patients with the same diagnosis, all except one had uncomplicated delivery and pregnancy. The one complication, intraventricular hemorrhage, occurred in a patient in whom an extracranial-intracranial bypass had not been performed.

The article is relevant and informative. It tells us that the natural history of moyamoya disease may be benign in some cases. Pregnancy, which places undue stress on the cerebral vessels because of blood volume changes, may be well tolerated, even without surgery. The one case of bleeding may have occurred even if the patient had already undergone bypass surgery. The many operations that are now performed to

relieve progression may be inappropriate until we know more about the natural history of this disorder. Our colleagues who find this disease in prevalence should organize a cooperative venture to answer the many questions that patients and families who face this disorder ask of their physicians.

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This is an excellent review of the literature concerning patients who either were diagnosed as having moyamoya disease because they became symptomatic during pregnancy or who had a diagnosis of moyamoya disease and then became pregnant. In a very logical fashion, the authors separate these two groups of patients.

The authors conclude that there is no evidence that pregnancy increases the risk of cerebrovascular accident or that bypass surgery decreases its risk. I think that their first conclusion is justified in that among the group of 30 patients who were known to have moyamoya disease and then went through pregnancy and delivery, there was only 1 with a poor outcome from bilateral ventricular hemorrhage at 30 weeks of gestation. Four patients had transient ischemic attacks or reversible ischemic neurological deficits, but they did well. The data supporting the second conclusion, that intracranial bypass does not make a difference, are not as conclusive, and I am not sure that a definite statement can be made. The authors also conclude that in patients with moyamoya disease, either cesarean section or vaginal delivery can be accomplished safely and that any method of anesthesia can be used provided appropriate precautions, particularly avoidance of hypocapnia, are taken. I think that their data justify this conclusion.

Even though the risk of a serious cerebrovascular accident during pregnancy is small, I am not sure that we can tell patients with moyamoya disease that pregnancy does not carry some risk. The second group of patients reviewed by the authors include 23 patients who became symptomatic, usually because of hemorrhage or ischemic events, during pregnancy. With the high frequency of moyamoya disease in Japan, it is difficult to know whether this means that pregnancy is risky for these patients. It does mean, however, that problems can develop during pregnancy and that these patients have to be particularly careful if they choose to become pregnant. The authors make a series of very sensible recommendations regarding how to guide these patients through pregnancy and delivery. We are thankful to these authors for providing us with an excellent summary of this topic and with very useful recommendations for a situation that, although very uncommon in the Western hemisphere, does present itself with sufficient frequency to make it important for all of us to be well informed on this topic.

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