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Peri-Operative Complications in Adult Moyamoya Disease

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Summary

The incidence and causes of peri-operative haemodynamic complications in adult Moyamoya disease were examined by reviewing 55 surgically treated adult patients. Ninety-nine craniotomies were performed in these patients, and eight peri-operative complications (four infarctions, two haemorrhagic infarctions and two reversible ischaemic neurological deficits without a new lesion) were seen. All of the eight haemodynamic complications arose in the initially affected hemispheres regardless of the side of operation. Some non-surgical haemodynamic risk factors, i.e., hypercapnia, hypocapnia and hypotension/hypovolaemia, were noted in all of the eight cases, although the statistical analysis could not clarify the relevance of such factors to peri-operative complications. Surgical factors which might be responsible for the complications were noted in three cases.

Sparing vital collateral vessels and minimum brain retraction as well as avoidance of non-surgical haemodynamic risk factors are considered to be essential to prevent peri-operative haemodynamic brain damage in adult Moyamoya disease.

Keywords: Moyamoya disease; adult; peri-operative haemodynamic complications; surgical factors; non-surgical factors.

Introduction

Moyamoya disease can be divided into two clinical entities, namely juvenile and adult types[7, 12, 14]. Although these two types show similar angiographic findings, it is not clear whether adult cases are only the extension or the continuity of juvenile cases or not. The juvenile type of Moyamoya disease is found after cerebral ischaemic attacks in almost all the cases during childhood. In contrast, the adult type often manifests intracranial bleeding and less frequent cerebral ischaemic attacks.

For Moyamoya disease associated with cerebral ischaemia, extracranial/intracranial (EC/IC) bypass has been done in the hope of preventing further ischaemic insults[1–4, 14]. Because of the disturbance of autoregulation under hypoperfusion states[8, 13], special care is necessary to prevent hypotension and hypocapnia during the peri-operative periods[5, 6, 9, 10]. In addition, we have also noted the potential hazard of peri-operative hypercapnia worsening the ischaemic conditions in juvenile cases (manuscript submitted).

In adult cases of Moyamoya disease, not only the EC/IC bypass but also evacuation of haematoma, clipping of associated aneurysms and other surgical procedures may become necessary. These operations should be done when there is cerebral hypoperfusion, narrowing of the major arteries or abnormal dilatation of the perforating vessels.

Table 1. *Clinical Profile of 55 Patients with Adult Moyamoya Disease*

Age at onset (yrs)	
Mean	37.3
Range	16–62
Male : Female	20 : 35
Manifestation	
Infarction	14
TIA	6
Haemorrhage	33
Incidental	2
Surgical procedure	
Direct bypass surgery	85
Indirect bypass surgery	1
Direct bypass surgery with another procedure	4[a]
Evacuation of haematoma	7
Resection of peripheral aneurysm	2

[a]Including clipping of the neck and coating of an aneurysm, a proximal clipping for distal aneurysm and a removal of meningioma.

In the present study, we have analysed 55 cases of surgically treated adult Moyamoya disease to clarify contributory factors of peri-operative complications.

Clinical Materials and Methods

We have performed 99 craniotomies on 55 adults (16 years old or more at onset) with Moyamoya disease in the past 15 years. Table 1 summarizes the clinical findings in these 55 patients.

Eight peri-operative haemodynamic complications (four infarctions, two haemorrhagic infarctions, and two reversible ischaemic neurological deficits (RIND) without a new lesion on neuro-imaging studies) were seen in eight patients. Postoperative transient ischaemic attacks (TIA) were excluded from this analysis.

Clinical records of these eight patients were analysed in detail with special reference to surgical procedures and peri-operative haemodynamic conditions, such as mean arterial blood pressure (MAP), blood gas data, as well as neurological signs and neuroradiological findings. Non-surgical haemodynamic factors were defined in the present study as hypercapnia ($PaCO_2 > 45$ torr), hypocapnia ($PaCO_2 < 35$ torr) and hypotension/hypovolaemia (MAP < 85% of the pre-operative MAP).

The data of these eight patients were compared with those of the 47 adult patients without such a complication.

Statistical analysis was performed using the Chi-square test.

Results

Features of the Patients with a Peri-Operative Complication

Table 2 summarizes the clinical data of the eight patients with a peri-operative haemodynamic complication. Diagnosis of Moyamoya disease was made after intracranial bleeding in four cases (two with thalamic and two with intraventricular haemorrhage) and after cerebral ischaemia in three cases (two infarctions and

Table 2. *Clinical Profile of Eight Patients with Peri-Operative Complications*

Case no.	Age (yrs/ sex)	Manifestation		Peri-operative complication			Probable cause of complication	
		Type	Affected hemisphere	Mode of operation	Type	Location of LDA/HDA	Non-surgical factor[a]	Surgical factor
1	23/F	infarction	right	lt. STA-MCA	infarction	rt. frontal	hypocapnia	none
2	29/M	incidental (head trauma)	none	rt. STA-MCA	infarction	lt. temporo-parietal	hypercapnia	none
3	38/M	thalamic haemorrhage	left	lt. STA-MCA, "neck" clipping of lt. P1–P2 aneurysm via subtemporal approach	infarction	lt. frontal	hypocapnia, hypercapnia, hypotension/ hypovolaemia	narrowing of the posterior communicating artery by the clip
4	41/F	intra-ventricular haemorrhage	left	resection of lt. distal aneurysm via transcortical approach	infarction	lt. parieto-occipital	hypotension/ hypovolaemia	retraction of the brain
5	18/M	infarction	right	rt. STA-MCA	haemorrhagic infarction	rt. parieto-occipital	hypercapnia	none
6	29/F	thalamic haemorrhage	left	lt. STA-MCA	haemorrhagic infarction	lt. fronto-parietal	hypercapnia	none
7	17/M	transient ischaemic attack	right	rt.STA-MCA	RIND corresponding to the operated hemisphere	none	hypercapnia, hypotension/ hypovolaemia	none
8	49/F	intra-ventricular haemorrhage	left	lt. STA-MCA, feeder clipping of lt. distal aneurysm via interhemispheric transcallosal approach	RIND corresponding to the operated hemisphere	none	hypocapnia	retraction of the brain

LDA low density area; *HDA* high density area; *F* female; *M* male; *STA-MCA* superficial temporal artery to middle cerebral artery anastomosis; *RIND* reversible ischaemic neurological deficit.

[a]See text for the definitions of hypercapnia, hypocapnia and hypotension/hypovolaemia.

one TIA); the remaining one case (Case 2) was diagnosed incidentally after head trauma.

EC/IC bypass surgery was done in five patients. In the other two patients, bypass surgery was done at the same time with aneurysmal clipping (neck clipping of a P_1–P_2 aneurysm and feeder clipping of a distal aneurysm). In the other patient, resection of a distal aneurysm of the lateral posterior choroidal artery was done.

In all of eight patients, haemodynamic complications arose in the initially affected hemispheres regardless of the side of operation. In Cases 1 and 2, a postoperative infarction arose in the contralateral hemispheres to the bypass surgery at the second operation. The first bypass surgery had been already done in these hemispheres, because cerebral blood flow (CBF) was predominantly reduced in these hemispheres before the first operation. In the remaining six patients, peri-operative haemodynamic complications occurred in the operated hemisphere, although no lesions were newly seen on the postoperative computerized tomography (CT) scans in the two cases of RIND (Cases 7 and 8).

Probable Causes of Peri-Operative Complications

The involvement of surgical factors in the complications was suspected in the three patients who underwent additional surgical procedures besides bypass surgery (Table 2). Narrowing of a vital collateral vessel was noted postoperatively in Case 3 and retraction of the brain was suggested to be one of the probable causes of the postoperative deterioration in Cases 4 and 8. Postoperative angiography confirmed the patency of EC/IC bypasses in all the patients, and surgical factors to which the complications might be attributable were not associated with these bypass surgeries.

In all of the eight patients, some non-surgical haemodynamic risk factors were seen with various combinations during the peri-operative periods. Hypercapnia, hypotension/hypovolaemia and hypocapnia were

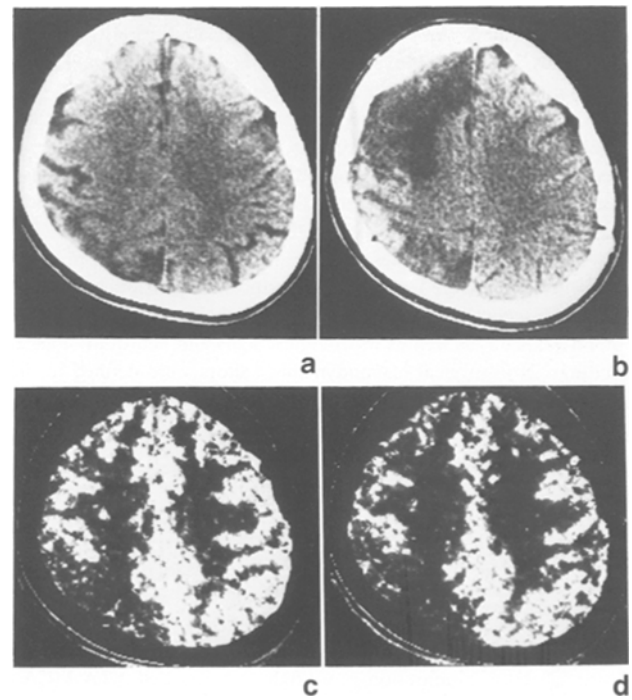

Fig. 1. Case 1. Pre-operative CT (a) demonstrating atrophy following infarction at the right parieto-occipital region. CT (b) obtained after the second operation (left STA-MCA anastomosis) revealed an infarction in the right fronto-parietal region. Cortical hyperdensity is seen in the right parietal region. Pre-operative Xe-CT images at rest (c) and after acetazolamide (Diamox) challenge (d). Cerebral blood flow (CBF) is shown to be decreased at rest in the right hemisphere, especially in the right perieto-occipital region corresponding to the infarction shown in pre-operative CT. CBF is further decreased after Diamox challenge in the fronto-parietal regions bilaterally but predominantly on the right

Table 3. *Peri-Operative Non-Surgical Haemodynamic Factors in Eight Adult Patients with Peri-operative Complications*

Peri-operative complications	Hypercapnia	Hypocapnia	Hypotension/ Hypovolaemia
Yes (n = 8)	5 (62.5%)	3 (37.5%)	3 (37.5%)
Infarction (n = 4)	2	2	2
Haemorrhagic infarction (n = 2)	2	0	0
RIND without LDA (n = 2)	1	1	1
No (n = 47)	22 (46.8%)	22 (46.8%)	16 (34.0%)
Total (n = 55)	27 (49.1%)	25 (45.5%)	19 (34.5%)

RIND reversible ischaemic neurological deficit; *LDA* low density area. See text for definitions of hypercapnia, hypocapnia and hypotension/hypovolaemia.

noted in five, three and three patients, respectively. Table 3 shows the relationship between the type of complication and these factors. There was no difference in the incidence of every factor between the patients with and without a complication, although both of the two patients with a haemorrhagic infarction had evidence of hypercapnia without any other factors.

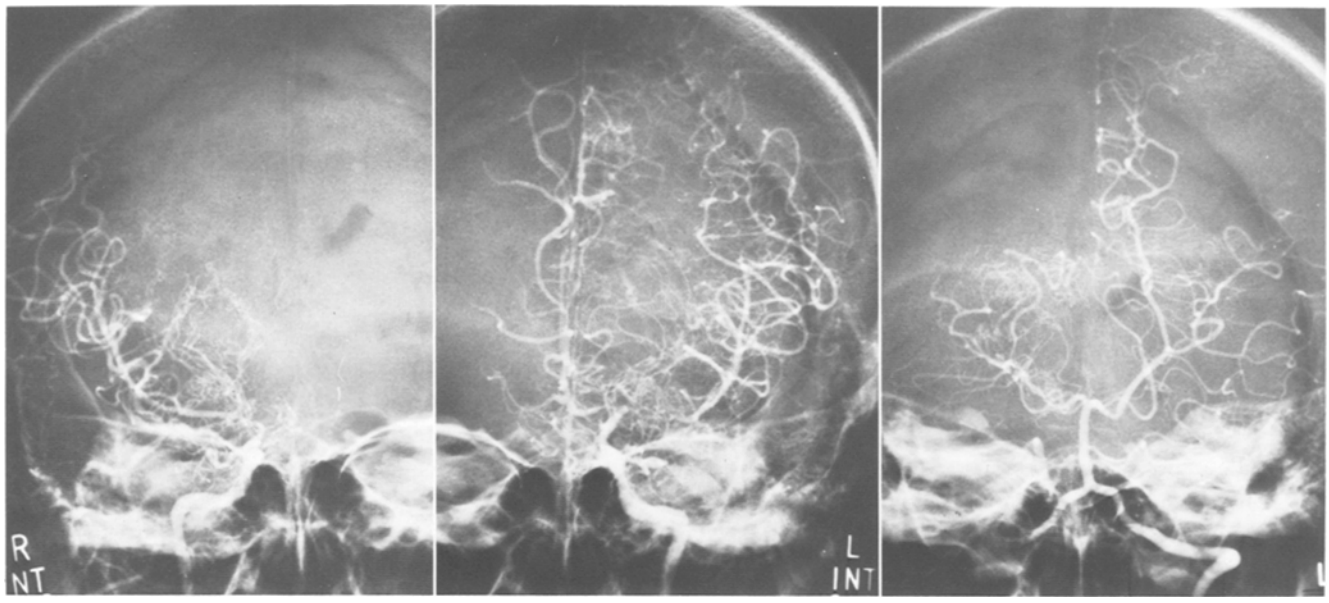

Fig. 2. Case 1. Antero-posterior views of right (left) and left (middle) CAG demonstrating bilateral stenoses of the internal carotid artery at its terminal portion and the origins of middle and anterior cerebral arteries. Left VAG (right) showing occlusion of the right posterior cerebral artery

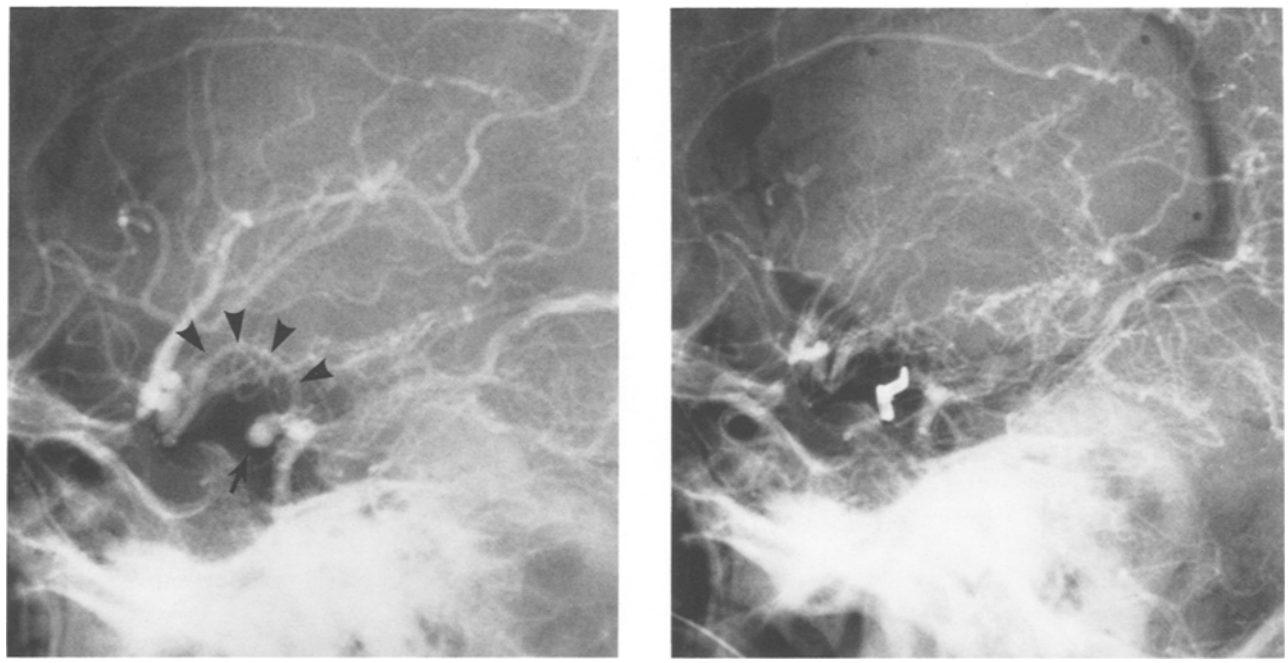

Fig. 3. Case 3. Lateral views of pre-operative (a) and postoperative (b) right VAG. A saccular aneurysm located at the left P 1–P 2 portion is seen (arrow). The left anterior and middle cerebral arteries are filled through the enlarged left posterior communicating artery (PCoA; arrowheads). Postoperatively, the aneurysm is not shown, but the left PCoA is remarkably stenotic; collateral flow to the anterior and middle cerebral arteries through the PCoA becomes poor

Illustrative Cases

Case 1

This 23-year-old woman developed a left hemiparesis with left homonymous hemianopsia. Her left hemispheres improved to nearly normal within four weeks. A CT scan (Fig. 1 a) demonstrated atrophy following infarction in the right occipital lobe. Cerebral angiography (Fig. 2) revealed bilateral narrowing of the internal carotid artery (ICA) at its terminal portion and the right posterior cerebral artery, indicating Moyamoya disease. A Xenon-CT scan (Xe-CT; Fig. 1 c) showed the hypoperfusion in the right hemisphere, especially in the occipital lobe corresponding to the infarction. The Xe-CT with acetazolamide (Diamox) challenge (Fig. 1 d) revealed the reduction of haemodynamic reserve in the fronto-parietal region bilaterally but predominantly on the right side.

She underwent right superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis. One month after the first bypass surgery, left STA-MCA anastomosis was done.

Postoperatively, she exhibited left hemiparesis. A CT (Fig. 1 b) demonstrated a new low density area (LDA) in the right fronto-parietal region in which reduction of haemodynamic reserve was indicated by the pre-operative Xe-CT study. She was discharged with mild weakness in her right fingers two months after the second operation. Postoperative angiography confirmed the bypasses to be patent. Hypocapnia (minimum $PaCO_2$ = 25.4 torr) was seen during her intra- and postoperative periods. No other risk factors were noted in her peri-operative clinical course.

Case 3

This 38-year-old man suddenly developed speech difficulty and right hemiparesis. A CT scan showed a haematoma in the left thalamus penetrating into the lateral ventricle. Cerebral angiography revealed narrowing of the right ICA at its terminal portion, occlusion of the left ICA at just distal to the origin of the ophthalmic artery and so-called "moyamoya vessels". A saccular aneurysm was also demonstrated at the left P_1–P_2 portion (Fig. 3 a).

Four months after the ictus, his hemiparesis recovered markedly and he underwent clipping of the neck of the aneurysm via the subtemporal approach. Left STA-MCA anastomosis was done at the same time.

Postoperatively, his hemiparesis deteriorated and a CT showed a large LDA in the left frontal lobe. Postoperative angiography (Fig. 3 b) revealed marked narrowing of the left posterior communicating artery (PCoA) and poor filling of the anterior circulation through the PCoA.

The infarction was considered to be caused by reduction of the blood flow through the PCoA, being a vital collateral to the infarcted area. In this case, intra-operative hypercapnia and hypotension/hypovolaemia were noted and hypocapnia was seen postoperatively. His hemiparesis did not improve in the follow-up period.

Discussion

Moyamoya disease is a clinical entity characterized by bilateral spontaneous occlusion of the ICA at its terminal portion[7, 11]. The age distribution at onset shows two peaks in the first and fourth decades[12, 14]. It is not clear whether these two types have the same pathogenesis and pathophysiology. It is also not clear whether decreased haemodynamic reserve or autoregulation affects the operative results in adult cases as in juvenile cases.

In contrast to juvenile cases, the majority of adult cases are associated with intracranial bleeding[7, 12, 14]. Cerebral ischaemic attacks are seen less frequently in adult cases than juvenile cases, even when they are presented by cerebral ischaemia. Therefore, the haemodynamic conditions in adult cases are considered to be more stable than those in juvenile patients. However, the incidence of peri-operative haemodynamic complications was 14.5% (8/55) in adult patients of our series, similar to that in juvenile patients (16.9%, 21/124) (manuscript submitted). Moreover, there was no difference in the incidence of such a complication between the haemorrhagic type and the ischaemic type.

There are two major points concerning the cause of peri-operative haemodynamic complications in adult Moyamoya disease. The first is the involvement of non-surgical haemodynamic factors[5, 8–10, 13]. In our series, although the statistical analysis could not clarify the relevance of such factors to haemodynamic complications, some risk factors were noted in all of the eight patients with a peri-operative complication. This indicates that these haemodynamic risk factors promote cerebral hypoperfusion in adult patients as well as in juvenile patients. Even if patients had no history of cerebral ischaemic attacks, their haemodynamic reserve is often reduced, as assessed by CBF studies. Moreover, a CBF study with Diamox challenge is useful for evaluating the risk of peri-operative complications (manuscript submitted). Therefore, an adequate CBF study should be done to the extent possible in all the patients with Moyamoya disease, although it may be difficult in the patients requiring urgent surgery for intracranial bleeding. Peri-operative management under normocapnia and normotension would be better to prevent peri-operative complications even in adult cases[10].

The other point is the influence of operative procedures. In our series, surgical factors that might be involved in the peri-operative complications were not seen with bypass surgeries. Sparing of vital collateral vessels is essential to prevent peri-operative ischaemic brain damage and to provide a better outcome. Minimum brain retraction is also important. In our two cases (Cases 4 and 8), a self-retaining retraction system was used and the retraction force was considered to be usual or relatively low in a retrospective review of the operation video. However, an infarction arose in the retracted area in Case 4 and postoperative neurological

deterioration continued for a month in Case 8. Brain tissue in Moyamoya disease would be in a hypoperfusion state and vulnerable to retraction or compression. In addition, when craniotomies are required for intracranial bleeding, revascularizations should be simultaneously done to improve such a hypoperfusion state.

In Moyamoya disease, in contrast with revascularizations for atherosclerotic occlusive cerebrovascular disease, EC/IC bypass operations could not achieve rapid improvement of the cerebral circulation. It takes several months to improve cerebral circulation after bypass surgery. Therefore, we should bear in mind that haemodynamic complications may arise within the area in which haemodynamic reserve was initially decreased, even where revascularization has been done.

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