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Childhood moyamoya disease before and after encephalo-duro-arterio-synangiosis: an angiographic study

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Summary. Encephalo-duro-arterio-synangiosis (EDAS) is a new surgical operation for childhood moyamoya disease, and its effects have been studied by comparing pre- and postoperative angiograms in 27 patients. The development of collaterals from the external carotid arterial system into the territory of the middle cerebral artery was excellent in 16 of 54 cerebral hemispheres after EDAS, good in 25, and poor in 13. The development of collaterals after EDAS increased as the stenotic process in the internal carotid artery on preoperative angiograms increased except in the most advanced cases, where it seemed to decrease in comparison with the group with middle grade stenosis. After EDAS, not only the superficial temporal artery, but also the adjacent middle meningeal artery participated in forming collateral pathways. Furthermore, decrease of abnormal net-like vessels was observed when there was good to excellent development of collateral vessels. Stenotic lesions in the internal carotid and posterior cerebral arteries were often seen to progress, indicative of the rapidly progressive nature of childhood moyamoya disease. These results appear to suggest that EDAS should be performed as early as possible in childhood moyamoya disease before the occurrence of an irreversible ischaemic state and/or permanent neurological defects.

Key words: Childhood moyamoya disease – Encephalo-duro-arterio-synangiosis – Cerebral angiography

Childhood moyamoya disease is a progressive, occlusive, cerebrovascular disease of unknown aetiology. It is most commonly seen in Japan [1–3], but cases have also been reported elsewhere [4, 5]. The peak incidence of this disease is in the first decade of life, and children afflicted at this age usually show evidence of ischaemia [3]. Angiography reveals bilateral stenosis or occlusion, primarily of the supraclinoid portion of the internal carotid artery (ICA), which extends to the proximal portions of the anterior and middle cerebral arteries (ACA, MCA) and parenchymal, leptomeningeal, and transdural collateral vessels which supply the ischaemic brain [6–8].

To minimize the progression of cerebral ischaemia in moymoya disease, several methods of surgical treatment have been introduced [9–12], one such being encephalo-duro-arterio-synangiosis (EDAS), a procedure for treating childhood moyamoya disease devised by Matsushima, one of the authors of this paper [13, 14]. This paper is an angiographic study of the effects of EDAS on childhood moyamoya disease, accomplished by comparing pre- and postoperative angiograms.

Materials and methods

From January 1981 to December 1988, 27 patients with childhood moyamoya disease, who were to be treated by bilateral EDAS, underwent pre- and postoperative angiography. As already reported [13], the EDAS operation diverts a scalp artery, usually the posterior branch of the superficial temporal artery (STA), under the skull and places it on the cortical surface for a certain length without interfering with the physiological blood flow through it. Sixteen of the 27 patients were male and 11 were female; their ages at the onset of symptoms and signs of moyamoya disease ranged from 1 to 10 years, average 5.4 years. Their ages at the time of preoperative angiography ranged from 1 to 20 years, average 8.3 years.

All underwent bilateral internal and external carotid arteriography, as well as uni- or bilateral vertebral arteriography, via a transfemoral catheter before and after EDAS. The delay between

Table 1. Classification of preoperative internal carotid angiograms according to the degree of stenosis and grade of abnormal net-like vessels (ANV) at the base of the brain

ICA stenosis		ANV from ICA[a]				ANV from PCA[a]			
Stage	No.	A	B	C	D	A	B	C	D
Stage 1	6	1	3	1	1	0	0	1	5
Stage 2	8	1	7	0	0	0	4	2	2
Stage 3	17	13	4	0	0	3	6	3	5
Stage 4	19	13	6	0	0	11	2	4	2
Stage 5	4	1	0	3	0	4	0	0	0
Total	54	29	20	4	1	18	12	10	14

[a] A, Marked; B, moderate; C, slight; D, none

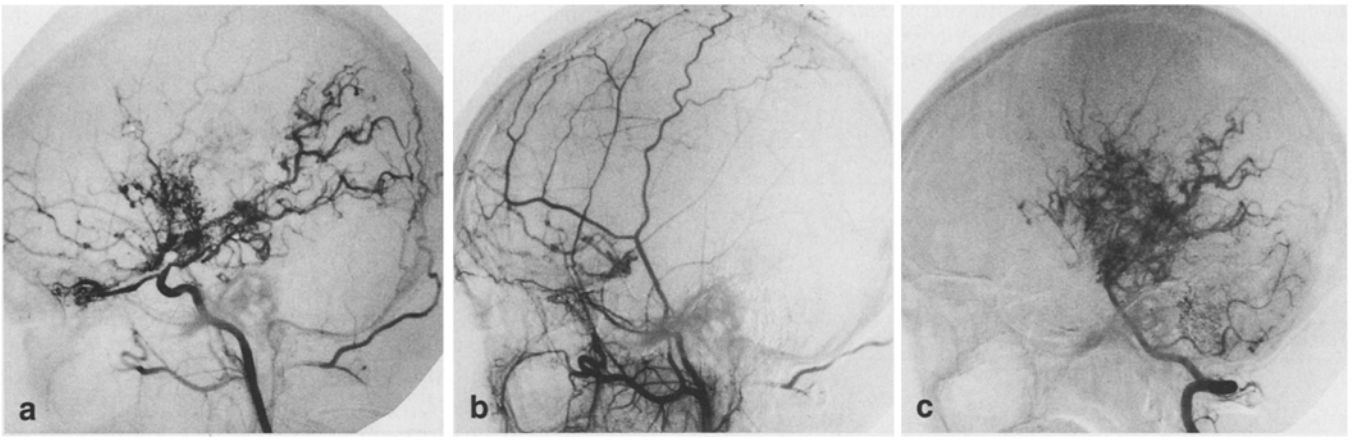

Fig. 1a–c. A 13-year-old girl. Preoperative angiogram. **a** Right internal carotid arteriogram the right internal carotid artery is occluded just above the origin of the posterior communicating artery, and the right middle and anterior cerebral arteries (MCA, ACA) are poorly visualized (stage 4). Prominent basal abnormal net-like vessels (ANV) are seen (grade A). **b** Right external carotid arteriogram. A little transdural collateral supply from the external carotid artery (ECA) is seen in the fronto-parietal region. **c** Left vertebral arteriogram. Prominent ANV are seen (grade A). The proximal posterior cerebral artery (PCA) is slightly stenosed on both sides

preoperative angiography and surgery ranged from 2 days to 2 months, average 20 days, and between surgery and postoperative angiography from 6 months to 3 years 2 months, average 11 months.

As reported previously [15], the internal carotid arteriograms were categorized, according to the degree of stenosis, into five stages: stage 1, slight to moderate stenosis of the ICA bifurcation (lumen ≥ 10%); stage 2, severe stenosis (lumen < 10%); stage 3, occlusion of ACA or MCA; stage 4, occlusion of ICA or ACA and MCA with partial retention of trunk of ACA or MCA; stage 5, occlusion of ICA or ACA and MCA with no filling of main trunk of either ACA or MCA.

The abnormal net-like vessels (ANV) at the base of the brain were divided into two groups: that fed by the ICA and its branches and that fed by the PCA and its branches. Further, the degree of ANV of each group was classified as A, marked; B, moderate; C, slight; and D, no visualization.

Postoperative development of collateral vessels from the external carotid arterial (ECA) system was assessed on the external carotid arteriograms following EDAS, and classified as excellent, good or poor. "Excellent" implied that all or most of the sylvian group of the MCA were well visualized via the newly developed collateral vessels from the ECA, "good" that several branches of the sylvian group were vis-

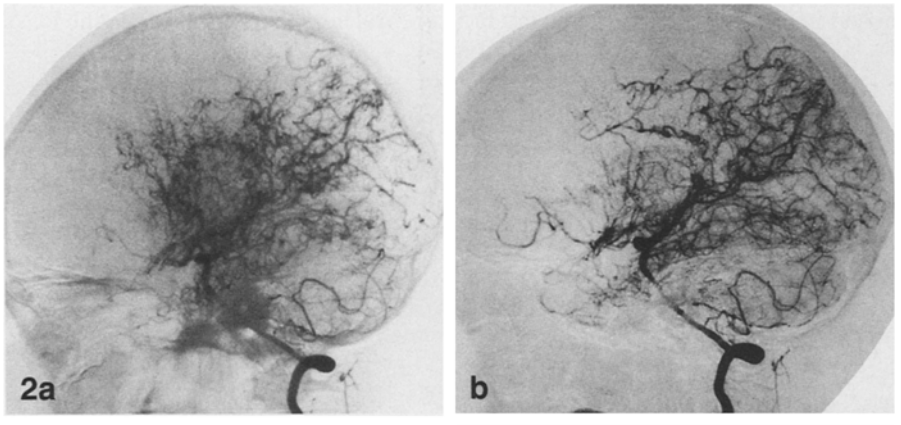

Fig. 2a, b. An 11-year-old boy
a Preoperative left vertebral arteriogram. Prominent ANV are opacified and dilated medullary arteries are seen in the deep parietal region (grade A). Prominent leptomeningeal anastomoses from the PCA to the posterior territory of the ACA are present
b Left vertebral arteriogram 7 months after EDAS. The ANV have regressed and the dilated deep medullary arteries have disappeared (grade B), while the leptomeningeal anastomoses have not changed significantly. External carotid angiography showed excellent development of collateral vessels

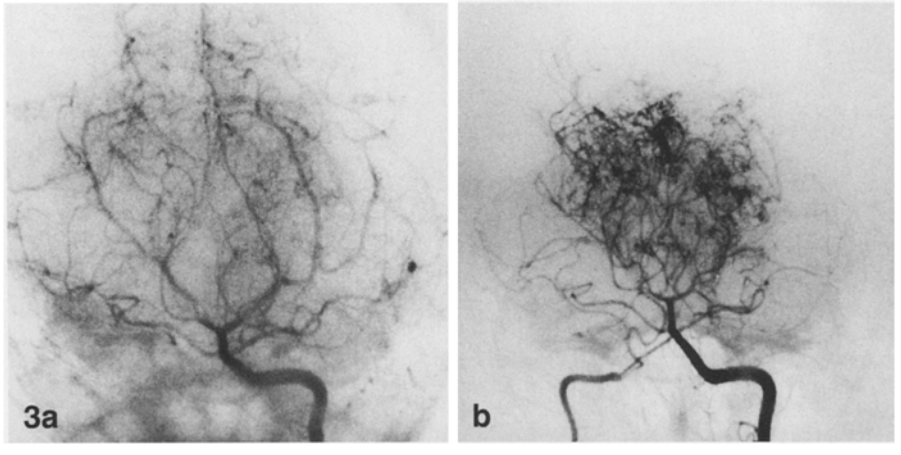

Fig. 3a, b. A 6-year-old girl
a Preoperative left vertebral arteriogram. The proximal PCA is stenosed on both sides, and slight ANV are seen (grade C)
b Left vertebral arteriogram 7 months after EDAS. The PCA is completely occluded proximally on both sides, showing progression of the stenotic lesion. There are marked ANV (grade A)

Table 2. Classification of preoperative internal carotid angiograms according to the degree of stenosis and development of collateral vessels from the external carotid artery (ECA) after encephalo-duro-arterio-synangiosis (EDAS)

ICA stenotic		Collaterals from ECA		
Stage	No.	Excellent	Good	Poor
Stage 1	6	1 (17%)	2 (33%)	3 (50%)
Stage 2	8	1 (13%)	5 (63%)	2 (25%)
Stage 3	17	5 (29%)	8 (47%)	4 (24%)
Stage 4	19	8 (42%)	9 (47%)	2 (11%)
Stage 5	4	1 (25%)	1 (25%)	2 (50%)
Total	54	16	25	13

ualized, and "poor" that few or no sylvian branches were visualized. Furthermore, the degree of dilatation of the STA and middle meningeal artery (MMA) following EDAS was estimated, and the ratio of the calibre of a given point in the stem of the artery on the postoperative and preoperative angiograms was calculated.

Results

Review of preoperative angiograms

The degree of stenosis in the ICA for the 54 cerebral hemispheres in 27 patients is shown in Table 1. The ANV at the base of the brain, fed by the ICA and PCA groups, increased with the severity of stenosis, but the ANV in the ICA group appeared to decrease in patients with stage 5 stenoses (Fig. 1). The PCA sent off leptomeningeal anastomoses to part of the ipsilateral ACA territory in 31 cerebral hemispheres, and to the ipsilateral MCA territory also in 31 (Fig. 2a). On preoperative angiograms, the peduncular segment of the PCA was stenosed in 15 of 54 hemispheres and occluded in 9 (Fig. 3a).

Development of collaterals from the ECA following EDAS

The development of collaterals from the ECA to the MCA territory was excellent in 16 of 54 cerebral hemispheres of patients who underwent EDAS, good in 25, and poor in 13 (Table 2). Development of collaterals also increased with the severity of the stenosis in the ICA on the preoperative angiograms, but seemed to decrease in stage 5, when compared with the moderate stages (Fig. 4).

It was found that the MMA and STA both participated in the formation of collateral pathways to the MCA territory in cases with good or excellent development of collateral vessels (Fig. 4b). In addition, the degree of dilatation of these arteries closely correlated to the development of collateral circulation (Table 3). There also was a significant difference in the calibre ratio of both the STA and the MMA on post- and preoperative angiograms when the patients with excellent or good results were compared with the poor result group ($P < 0.01$, by Student's t-test). Development of arteriovenous fistulae or communications between the STA or MMA and cortical veins was not observed following EDAS in any patient.

Changes in the ANV at the base of the brain

The ANV fed by the ICA group decreased following EDAS in 13 of the 54 cerebral hemispheres (Fig. 4a); 12 of these showed good or excellent development of collateral vessels (Table 4). The ANV fed by the PCA group was also seen to decrease in 12 cerebral hemispheres, and 11 of these showed good or excellent development of the collateral vessels (Figs. 2, 4c). In both groups there was a significant difference in the frequency of decrease of the ANV between the excellent and poor result groups ($P < 0.05$, by Fisher's exact probability test).

In contrast, no definite decrease was seen in the leptomeningeal anastomoses from the PCA to the ACA or MCA, except in 5 cerebral hemispheres in which the PCA was occluded (Fig. 2).

Progression of stenosis

Stenosis in the ICA was seen to have progressed on 12 of the 54 cerebral hemispheres as compared with the preoperative angiograms, reflecting the progressive nature of this disease over a relatively short interval (average

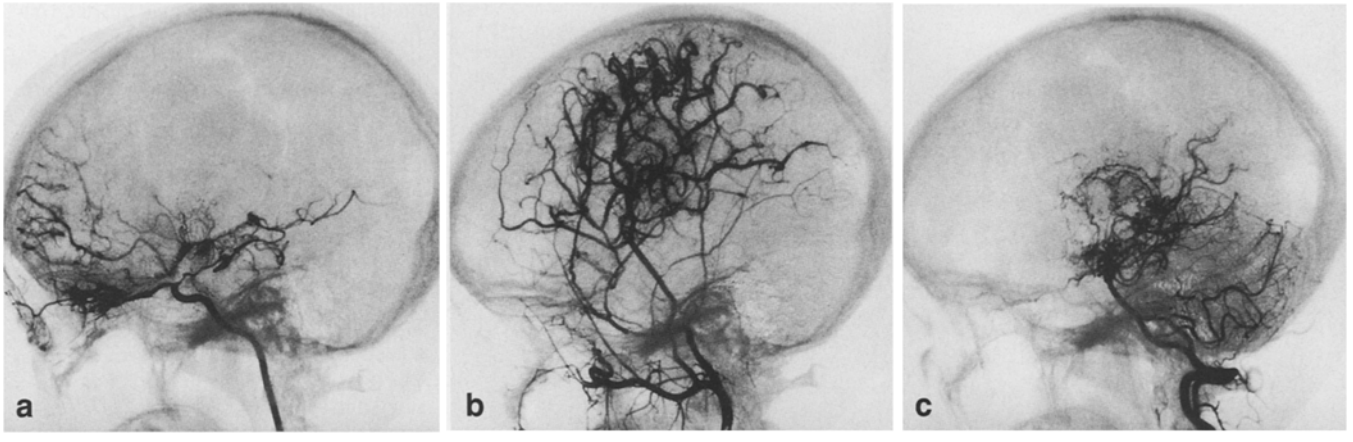

Fig. 4a–c. Same case as in Fig. 1. Angiogram 11 months after encephalo-duro-arterio-synangiosis (EDAS). **a** Right internal carotid arteriogram. The ANV have regressed (grade B); f. Fig. 1a. **b** Right external carotid arteriogram. The posterior branches of the superficial temporal artery and the middle meningeal artery are enlarged and almost all branches of the MCA are well seen (grade "excellent"). **c** Left vertebral arteriogram. The ANV have regressed (grade B); f. Fig. 1c

Table 3. Collateral vessels from the ECA after EDAS and dilatation of superficial temporal artery (STA) and middle meningeal artery (MMA)

Collateral vessels from ECA		Ratio of post/preoperative calibre (mean ± SD)	
Grade	No.	STA	MMA
Excellent	16	1.89 ± 0.33*	1.85 ± 0.29*
Good	25	1.72 ± 0.43*	1.68 ± 0.47*
Poor	13	1.12 ± 0.18	1.06 ± 0.16

* Significantly different from the grade of "poor" ($P < 0.01$, by Student's t-test)

Table 4. Collateral vessels from the ECA after EDAS and change in the ANV at the base of the brain

Collateral vessels from ECA		ANV from ICA		ANV from PCA	
Grade	No.	Decrease	Increase	Decrease	Increase
Excellent	16	7 (44%)*	0 (0%)	7 (44%)*	1 (6%)
Good	25	5 (20%)	1 (4%)	4 (16%)	3 (12%)
Poor	13	1 (8%)	1 (8%)	1 (8%)	1 (8%)
	54	13	2	12	5

* Significantly different from the grade of "poor" ($P < 0.05$, by Fisher's exact probability test)

11 months): in 3 cerebral hemispheres from stages 1 to 2; in 1 from stages 1 to 3; in 1 from stages 2 to 4; in 1 from stages 2 to 5; in 5 from stage 3 to 4; and in 1 from stages 3 to 5.

Stenosis of the PCA had also progressed in 8 of the 54 cerebral hemispheres: in 5 from normal to stenotic, and in 3 from stenosis to occlusion (Fig. 3). In 5 of these, leptomeningeal anastomoses had also decreased markedly.

Discussion

Moyamoya disease is an occlusive cerebrovascular disease, and affected individuals usually show evidence of ischaemic problems in childhood [1–3]. As the occlusive lesions are confined to the distal ICA and the proximal portions of the cerebral arteries, the most logical approach to treatment appears to construct anastomotic channels to increase the blood supply to the brain [9–12]. EDAS has provided satisfactory clinical results, with the disappearance of or improvement in symptoms and signs [13, 14].

Our data demonstrate that, following EDAS, the development of collateral vessels from the ECA to the MCA territory increased with the severity of the stenosis of the ICA on preoperative angiograms but that, in the most advanced stage, the development of collateral supply was less marked. EDAS differs from STA-MCA anastomosis in being an indirect ECA-ICA bypass in which transdural anastomoses form gradually [13]. These collateral vessels may develop better in a brain which undergoes progressively more ischaemic but still reversible states. Our data suggest that in stages 1 to 4 the brain is in an increasingly ischaemic but still reversible state, and that when it reaches stage 5, it becomes irreversibly ischaemic. Indeed, we found that there was no development of collateral vessels from the ECA after EDAS in areas which were of low density on CT, suggesting infarction. This suggests that EDAS for childhood moyamoya disease should be performed before an irreversible ischaemic damage supervenes.

The arteries feeding the collateral pathways following EDAS were derived from both the surgically treated STA and the adjacent MMA; in most cases the MMA dilated significantly and sent off numerous collateral vessels, indicating that, by enabling this opening of double channels from the ECA to the ischaemic brain, EDAS may significantly benefit collateral circulation. Ausman et al. [16] have reported a case in which multiple spontaneous anastomoses between the dural and cortical arteries were seen after an STA-MCA anastomosis. However, in childhood moyamoya disease after EDAS, the MMA seemed to play a much more important role in cerebral revascularization.

The ANV fed by the ICA regressed in many patients who showed good or excellent formation of collateral vessels following EDAS. In some the ANV fed by the PCA also regressed, when good or excellent formation of collateral vessels was achieved. A decrease in ANV following EDAS may be related to a decrease in a requirement for collateral flow to the cerebral cortex via them because of increased collateral circulation from the ECA. Takeuchi et al. [17] also reported decrease of ANV in moyamoya disease after encephalo-myo-synangiosis, another form of ECA-ICA bypass. This decrease may be of some benefit in preventing cerebral haemorrhage from these abnormal vessels. In contrast to the ANV, the leptomeningeal anastomoses of the PCA did not show a significant change, other than those cases in which stenosis in the PCA progressed, possibly due to the fact that these anastomoses mainly supply the territory of the ACA.

We demonstrated progression of the stenotic process over a relatively short interval in both the ICA and the PCA, indicating the rapidly progressive nature of childhood moyamoya disease. The leptomeningeal anastomoses from the PCA decreased in patients showing progression of stenosis in the PCA. Satoh et al. [15] have shown that the stenosis in the PCA progresses pari passu with that in the ICA. As the PCA is the most prominent collateral channel to the ICA, its progressive stenosis may lead to severe, irreversible ischaemia of the brain [18, 19]. Once complete infarction in the brain has occurred, there may be no development of collateral vessels from the ECA after EDAS. Our study thus suggests that EDAS for childhood moyamoya disease should be performed as early as possible, so as to prevent the development of irreversible ischaemia and/or permanent neurological defects.

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