The Specificity of the Collaterals to the Brain Through the Study and Surgical Treatment of Moyamoya Disease


SUMMARY Moyamoya disease presents clinically as chronic progressive ischemia in the young brain. The brain is surrounded by concentric collateral networks but all of these networks are not available as collaterals in the early stage of cerebral ischemia. The anatomical characteristics precluding their early use include the presence of the watery layer of subarachnoid fluid between the cortical and dural vessels and of a closed bony box intervening between the dural and scalp arterial networks. These barriers isolate the brain from the abundant blood flow of the external carotid system as if they were the moat (the subarachnoid fluid layer) and the walls (the skull) of a castle.

Based on these concepts, we have developed a surgical procedure, the encephalo-duro-arterio-synangiosis to treat moyamoya disease in children. This operation surmounts the above mentioned two obstacles to collateral formation to the brain by perforating the castle wall and bridging the moat by granulation tissue, without injuring the collaterals which are already formed.

This procedure was performed on 70 sides in 38 pediatric moyamoya patients. Revascularisation of the brain was obtained in 100 percent of the cases with varying improvement in the symptoms.

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MOYAMOYA DISEASE provokes chronic progressive stenosis or occlusion of unknown cause starting in the periphery of both internal carotid arteries. This disease is thus a model of chronic ischemia in the brain. Chronological angiographic observations in patients with this disease reveal the dynamics of formation of the collaterals to the brain in association with this chronic cerebral ischemia. They reveal the specificity of the cranial cerebral vascular architecture.

Collateral Systems to the Brain

The following is a suggested nomenclature of the collaterals and reserve collaterals to the brain (fig. 1).

1. Anastomosis Intracerebralis (A System)

There are two sets of perforating arteries to the brain, one from the base of the brain and another from the surface of the brain. They anastomose with each other at the external angle of the lateral ventricle. This anastomosis has been clarified by many investigators, though it can not be noted on usual cerebral angiograms. This anastomosis is also seen by cerebral angiography in diseases other than moyamoya under some conditions, and Takeuchi appropriately calls it the moyamoya phenomenon. Figure 2 shows the well-developed intracerebral anastomosis (A system), called moyamoya vessels, in a moyamoya patient.

2. Basal Communications (B System)

The B system includes the circle of Willis and persistent carotid-basilar anastomoses. The primary lesion in moyamoya disease is in the circle of Willis, and the disease is also called “Willis’ arterial ring occlusion disease” in Japan, so the disease can be defined as a disease of B system insufficiency.

3. Cortical Leptomeningeal Anastomosis (C System)

This is an end-to-end anastomoses of 200-600 μ size between the principal cerebral arteries on the surface of the brain. There are extreme individual differences in these anastomoses and the variation of symptoms in different patients with equivalent moyamoya, emboli, or cerebral vasospasms seems to be related to these individual differences.

4. Dural Networks (D System)

The dural arteries are anastomosed with each other as if the brain were covered with a mesh hat composed of the dural arteries. Therefore, if there were no subarachnoid CSF layer, and if the dural networks were in direct physical contact with the brain, this system could be utilized as a collateral blood source immediately after ischemia occurred in the brain. However, as the CSF layer usually separates the two systems, a rapid anastomosis cannot be formed between them. The dural network eventually forms an effective anastomosis with the C system via transdural anastomoses. In the natural progress of moyamoya disease, several months or years are required before these anastomoses are confirmable by cerebral angiography. Surgical procedures will obviate this delay.

5. Extracranial Networks (E System)

The blood flow in the vascular networks of the scalp and pericranial muscles is abundant. If the cranium were not a hard closed box, this network would be an ideal source of blood supply for the cerebrum. Unless the cranium is perforated by surgical procedures, it will take several months or years before this system can provide sufficient collaterals to the ischemic brain.

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CSF, Skull -- Scalp Cranium Transdural anastomosis Cerebrum Ventricle Dura mater Cerebral surface Subarachnoid space (F)

FIGURE 1. Specificity of the collaterals to the brain. There are concentric layers of collaterals from A to G systems: A = Anastomosis intracerebralis; B = Basal communications; C = Cortical leptomeningeal anastomosis; D = Dural networks; E = Extracranial networks; F = Functional collaterals; G = Ground communications. There are also a subarachnoid fluid layer between the C and D systems and the solid closed box of the cranium between the D and E systems. Both prevent the D and E systems being easily utilized as collaterals.

6. Transdural Anastomosis (T System)

In order for the E and D systems to become effective as collateral pathways, they must anastomose with the C system. This bridging anastomosis is the so-called transdural anastomosis. There has been no detailed report on the development of this system. However, when this anastomosis develops spontaneously in cases of advanced moyamoya disease, it probably arises at the regions where some soft tissue intervene between the C system and the D and E systems: where the bridging veins enter the dural sinuses; where there are arachnoid villi; or where arteries or nerves bridge the dura mater and the brain.

In clinical practice, these anastomoses exist chiefly near and along the midline as confirmed by cerebral angiography, and in the ethmoidal region of the base of the brain. Figure 3 shows these transdural anastomoses with the D and E systems acting as collaterals in advanced moyamoya disease.

The cranium, as a hard closed box, intervenes between the E and D systems, and a fluid layer of CSF intervenes between the C and D systems: like layers in a sandwich. Moyamoya disease, with its particular symptomatology and characteristic angiograms, is nothing but the product of this specific craniocerebral vascular architecture.

Dynamics of Formation of Collaterals in Moyamoya Disease

It is helpful to follow the natural progress of pediatric moyamoya disease, a natural model of chronic progressive ischemia in the young brain, utilizing this nomenclature.

As Suzuki and Kodama have reported, dynamic changes are observed in serial angiograms made on pediatric patients suffering from moyamoya disease. The staging of the disease based upon the angiographic findings conforms well to these observations and the hypothesis that intracranial collaterals are gradually replaced in the progression of the disease by extracranial collaterals which have greater capacity. The state in which sufficient blood supply is obtained through the external carotid system is the state in which so-called natural healing has occurred. The dynamics of formation of the collaterals in moyamoya disease viewed in relation to the above described nomenclature of collaterals and specific anatomical characteristics are as follows:

The primary lesion in moyamoya disease, of unknown cause, slowly and progressively occludes the B system starting at the periphery of the bilateral internal carotid artery (Stage I of Suzuki). In proportion to the blood flow decrease due to this stenosis, the collateral systems develop or grow successively, in the order of ease of their development.

With the advancing B system insufficiency, both the A and C systems are mobilized almost simultaneously and compensate for the B system insufficiency. Any failure in this compensation is expressed as an ischemic episode common to the moyamoya patient. The C system is readily overlooked as it consists of nothing more than the cortical arteries visible on usual cerebral angiograms. In contrast, the A system shows a peculiar picture never present in normal cerebral angiograms.

FIGURE 2. Anastomosis intracerebralis (A system) found well developed in a moyamoya patient.
SPECIFICITY OF COLLATERALS TO THE BRAIN/Matsushima and Inaba

(Stage II of Suzuki). The distinctive A system attracted early attention in the early descriptions of this disease and has been described as moyamoya vessels, an abnormal vascular network at the base of the brain. As the oligemia becomes severe, the A system becomes quite marked (Stage III of Suzuki, Figure 2). When this ischemic state persists, the blood flow in the minute arterioles in such soft tissues as those in the fibrotic suture sites or at the site of the emissary vein, becomes increased between the D and E systems. The blood flow in the small arterioles accompanying the bridging veins, arachnoid villi, cranial nerves and arteries, which physically connect the dura mater and the brain, is increased between the D and C systems. The caliber of these arterioles increases gradually, and finally the so-called transdural anastomoses, called vault moyamoya or ethmoidal moyamoya by Suzuki et al begin to be apparent in the angiograms. When the transdural anastomosis between the C system and the D and E systems increases with time, the blood flow through the external carotid system increases. Consequently, the A system or the moyamoya vessels lose their role as collaterals to the brain and gradually becomes invisible by cerebral angiography (Stage IV of Suzuki).

(a) Further unfavorable conditions should not be added to the ischemic brain.
(b) The effective collaterals already formed and the reserve collaterals should not be damaged.
(c) The D and E systems with abundant blood flow and the C system should be anastomosed, by surmounting the two intervening obstacles.

A surgical procedure has been developed which is believed to satisfy all of these conditions and is called encephalo-duro-arterio-synangiosis (abbreviated as EDAS). This procedure (fig. 4) places the scalp artery, which is the principal artery of the E system, directly on to the surface of the brain by fixing the exposed scalp artery to the linear incisional edge of the dura mater through a narrow craniotomy. For easier handling of the donor scalp artery, the scalp artery is used with a narrow strip of galea attached, using the galeal strip to fix the donor artery to the dura mater. This operation is designed to promote formation of spontaneous transdural anastomoses. Linear incisions are used to the skin and to the dura mater to minimize the damage to the E and D systems. The donor artery is not cut at any portion and the blood flow in this artery

Rational Surgical Procedure To Treat Moyamoya Disease

From the hypothesis stated above, the ideal surgical procedure to treat moyamoya disease should fulfill the following conditions;
is maintained throughout the procedure, leaving all existing transdural anastomoses to function during and after the procedure.

**Operative Results of EDAS**

We have performed 70 EDAS on 38 pediatric moyamoya patients and, unlike other surgical techniques, have not observed any deterioration as a result of this operative procedure. Follow-up angiograms on these patients have shown marked dilatation of the dural arteries at 1-3 months after the operation and gradual dilatation of the donor artery associated with the revascularisation of the brain through the donor artery in almost all the cases at 6 months after the operation (table 1). Almost simultaneously the moyamoya vessels observed at the base of the brain have begun to decrease (fig. 5) and have completely disappeared in some cases.\(^1\)

Ischemic attacks seen in pediatric moyamoya patients such as TIA, seizures, and headache have disappeared in all cases within 21 months at longest (11.5 months in an average). EEG improvement has been seen in every case. Some of the existing motor deficits improved, but not much improvement of intellectual function has been obtained.

Figure 6 shows the external carotid angiograms of a moyamoya patient (Case 8 in Table 1) in whom the operation was performed 8 months previously. The whole hemisphere is visualized through the parietal branch of the STA, which is the donor artery. The type of revascularisation illustrated in Figure 6 occurred in each instance (table 1), and the symptoms subsided accordingly. Further neurological deterioration did not occur in any of the patients.

**Discussion and Conclusion**

Several indirect EC/IC anastomoses have been introduced in the treatment of moyamoya disease. These

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### Table 1 Preoperative and Postoperative Data Seen in 10 Consecutive Cases of Moyamoya Disease Who Underwent EDAS

<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Sex</th>
<th>Age at onset</th>
<th>Interval, onset to EDAS</th>
<th>Side</th>
<th>Interval after EDAS</th>
<th>Dilation of dural arteries</th>
<th>Brain revascularisation</th>
<th>Dilation of scalp artery</th>
<th>Decrease in moyamoya vessels</th>
<th>Brief descriptions of neurological changes</th>
</tr>
</thead>
</table>
| 1   | 12y 1m | ♂ | 6 y 10 m | 1 y         | rt   | 54 m         | +++ | +++ | +++ | +++ | + | Weakness attacks gone  
|     |       |    |             |             |      |               |              |     |           |                          | Lt. hand fingers movements improved  
|     |       |    |             |             |      |               |              |     |           |                          | Restlessness gone  
|     |       |    |             |             |      |               |              |     |           |                          | School achievements improved  
| 2   | 14y 9m | ♂ | 10 y 2 m | 5 m         | rt   | 46 m         | +++ | +++ | +++ | +++ | + | Rt. grasping power increased  
|     |       |    |             |             |      |               |              |     |           |                          | Weakness while running gone  
|     |       |    |             |             |      |               |              |     |           |                          | Weakness attacks gone  
|     |       |    |             |             |      |               |              |     |           |                          | Activity increased  
| 3   | 9y   | ♂ | 5 y 5 m   | 1 y 7 m  | rt   | 17 m         | +++ | + | +++ | +++ | + | Weakness attacks gone  
|     |       |    |             |             |      |               |              |     |           |                          | Activity increased  
| 4   | 9y   | ♂ | 5 y 3 m   | 1 y 7 m  | rt   | 6 m          | +++ | + | + | + | + | Weakness attacks gone  
|     |       |    |             |             |      |               |              |     |           |                          | Activity increased  
| 5   | 10y 8m | ♂ | 6 y 2 m   | 2 y 4 m  | rt   | 10 m         | +++ | + | +++ | +++ | + | Headache and TIA attacks gone  
|     |       |    |             |             |      |               |              |     |           |                          | Activity increased  
| 6   | 18y 4m | ♂ | 4 y 11 m  | 1y 3 m  | rt   | 19 m         | +++ | + | +++ | +++ | + | Headache and TIA attacks gone  
|     |       |    |             |             |      |               |              |     |           |                          | Activity increased  
| 7   | 11y 4m | ♂ | 3 y 6 m   | 9 m       | rt   | 8 m          | +++ | + | + | + | + | Headache gone  
|     |       |    |             |             |      |               |              |     |           |                          | Weakness attacks gone  
|     |       |    |             |             |      |               |              |     |           |                          | School achievements improved  
| 8   | 11y 4m | ♂ | 7 y 8 m   | 9 m       | rt   | 20 m         | +++ | + | + | + | + | Headache gone  
|     |       |    |             |             |      |               |              |     |           |                          | Weakness attacks gone  
|     |       |    |             |             |      |               |              |     |           |                          | School achievements improved  
| 9   | 10y 4m | ♂ | 5 y 8 m   | 2 y 10 m | rt   | 19 m         | +++ | + | + | + | + | Headache gone  
|     |       |    |             |             |      |               |              |     |           |                          | Activity increased much  
|     |       |    |             |             |      |               |              |     |           |                          | Weakness attacks gone  
| 10  | 10y 7m | ♂ | 6 y 4 m   | 2 y 4 m  | rt   | 6 m          | + | + | + | + | + | Weakness attacks gone  
|     |       |    |             |             |      |               |              |     |           |                          | Activity increased much  
|     |       |    |             |             |      |               |              |     |           |                          | No attacks even with Marathon  

are encephalo-myosynangiosis,\textsuperscript{19} encephalo-arteriosynangiosis,\textsuperscript{20} omental transplantation.\textsuperscript{21} All have proved to be more or less effective for the treatment of moyamoya disease by inducing spontaneous anastomoses between the arterial systems of the external and the internal carotid. Thus, the presence of an angiogenetic factor was postulated in moyamoya disease.\textsuperscript{19} There may be such a substance which promotes the spontaneous anastomoses between intracranial and extracranial arteries, but if it exists it may be a substance from ischemic brain and not specific to moyamoya brain. Physically surmounting the two obstacles to the formation of adequate collaterals from the external carotid system may contribute much more to the formation of the spontaneous EC/IC anastomosis. Clinical observation appear to conform with the validity of the concept that there are collateral vascular systems in the shape of concentric layers in and around the brain, and that if chronic cerebrovascular ischemia occurs, these collaterals will be utilized in the order of the ease of their access. Due to the specific anatomy of the head, only intracranial collaterals are utilized at first, but if
Figure 6. A selective external carotid angiogram of a moyamoya patient, taken 8 months after the encephalo-duro-arterio-synangiosis. Total hemisphere is visualized via the parietal branch of left STA, the donor artery (arrow).

the specific anatomical barriers are surmounted by time or by surgical procedures, the collaterals are utilized in the order of their effectiveness and the external carotid system with its greater collateral capacity predominates. As John Hunter said "blood goes where it is needed" and we would like to add "if there are no obstacles."

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