

Continuing Education Column

모야모야병의 뇌영상 진단과 치료

Neuroimaging Diagnosis and Treatment of Moyamoya Disease

300

Chang Wan Oh, M.D.
 Department of Neurosurgery
 Seoul National University College of Medicine, Bundang Hospital
 E - mail : wanoh@snu.ac.kr

Abstract

Moyamoya disease is characterized by bilateral progressive obliteration of the terminal internal carotid arteries. It is relatively common in Asian populations and is found more frequently in children, who present with symptoms and signs of cerebral ischemia, often provoked by hyperventilation during crying or eating hot food. In adult age, more patients present with intracranial hemorrhage. To differentiate moyamoya disease from other cerebrovascular diseases, the diagnosis has been defined according to a guideline provided by the 'Research Committee on Spontaneous Occlusion of the Circle of Willis'. Bilateral stenosis and/or occlusion with moyamoya vessels documented by cerebral angiography or MRA/MRI are the key points in diagnosis. Since its etiology is still unknown, prevention of progressive cerebral ischemic insult is the current target of treatment. Revascularization surgery, such as indirect or direct extracranial - intracranial bypass surgery can fulfill this goal. Early detection and treatment, especially in childhood, is most important for the better prognosis of this disease.

Keywords : Moyamoya disease; Definite moyamoya; Probable moyamoya; Direct revascularization surgery; Indirect revascularization surgery

(moyamoya disease, MMD)

1950

(1, 2), 1963 Jiro Suzuki 가 6

(3, 4).

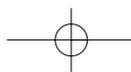
(spontaneous occlusion of the circle of Willis) ,

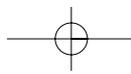
1967 Jiro Suzuki 가 ' ,

1969

(5)

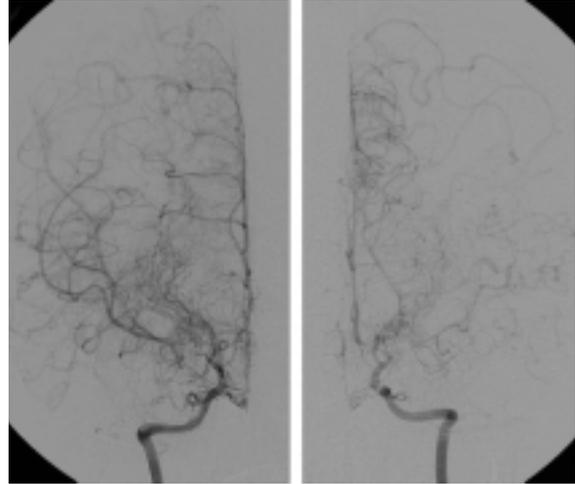
가





Continuing Education Column

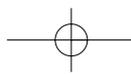
가 ,
 , (MRI),
 (MRA)
 1977
 (Research Committee on Spontaneous Occlusion of
 the Circle of Willis) ' 1978
 , 1988 1995
 (6). 가

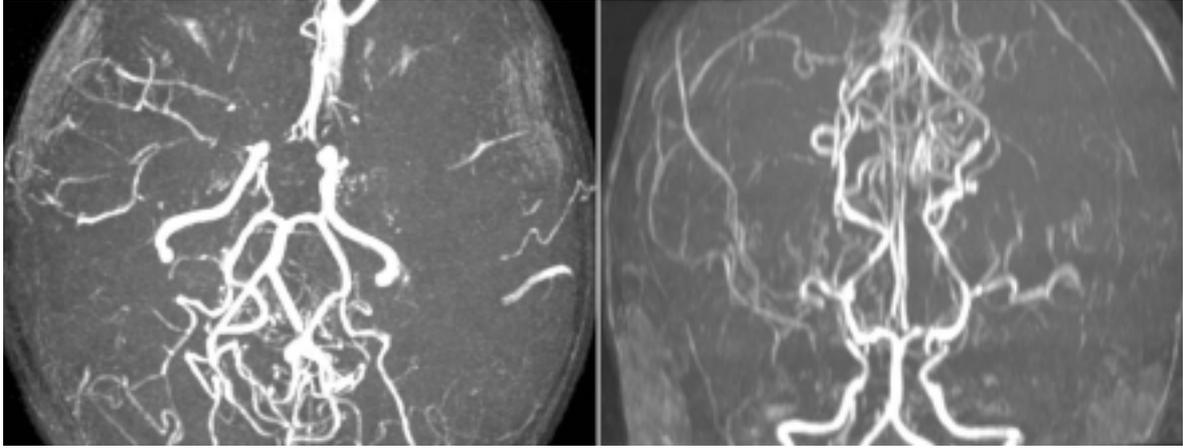
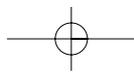


1.

MRI, MRA
 가 ,
 (signal void)
 (3)
 1.
 1) (1)
 (1)
 (2)
 (arterial phase) (abnor-
 mal vascular network)
 (3)
 2) MRI MRA (2, 3)
 (1) MRA
 (2) MRI

(signal void)
 가
 (3)
 3)
 , 가 , , ,
 , Recklinghausen , ,
 4)
 (1) (intimal
 thickening)
 (2) -
 (fibrocellular thickening),
 (waving),
 (3)





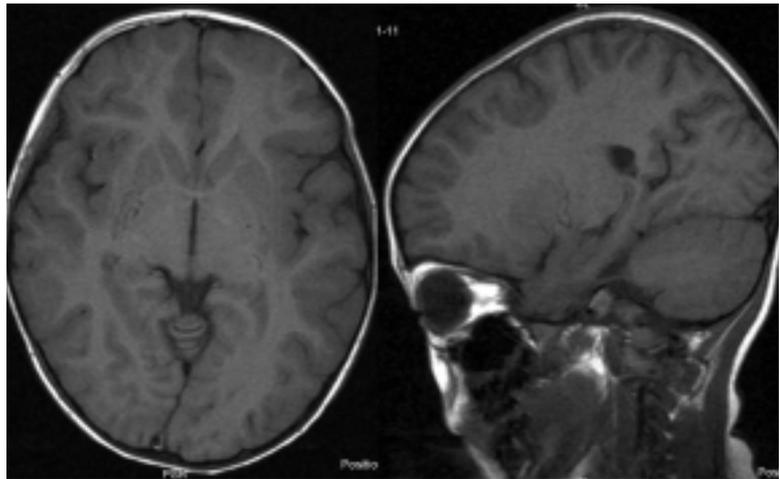
2. MRA

(4) (pia mater)

2.

1) (確診性)
(Definite Cases)

MRI MRA



3. MRI

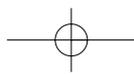
MRI MRA

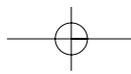
2) (疑診性) (Probable Cases)

(screening)

가

(probable moyamoya disease)'





Continuing Education Column

1. Suzuki stages

Suzuki stage
I
II 가
III 가
IV (ethmoidal moyamoya vessels) 가
V 가
VI

3. (Suzuki Stages)
Jiro Suzuki (5)

6 (stages)
(1)(4).

1 6

4.

MRI CT가 PET(positron emission tomography) rCBF(regional cerebral blood flow), rCMRO2 (regional cerebral metabolic rate of oxygen), rOEF(regional oxygen extraction fraction)

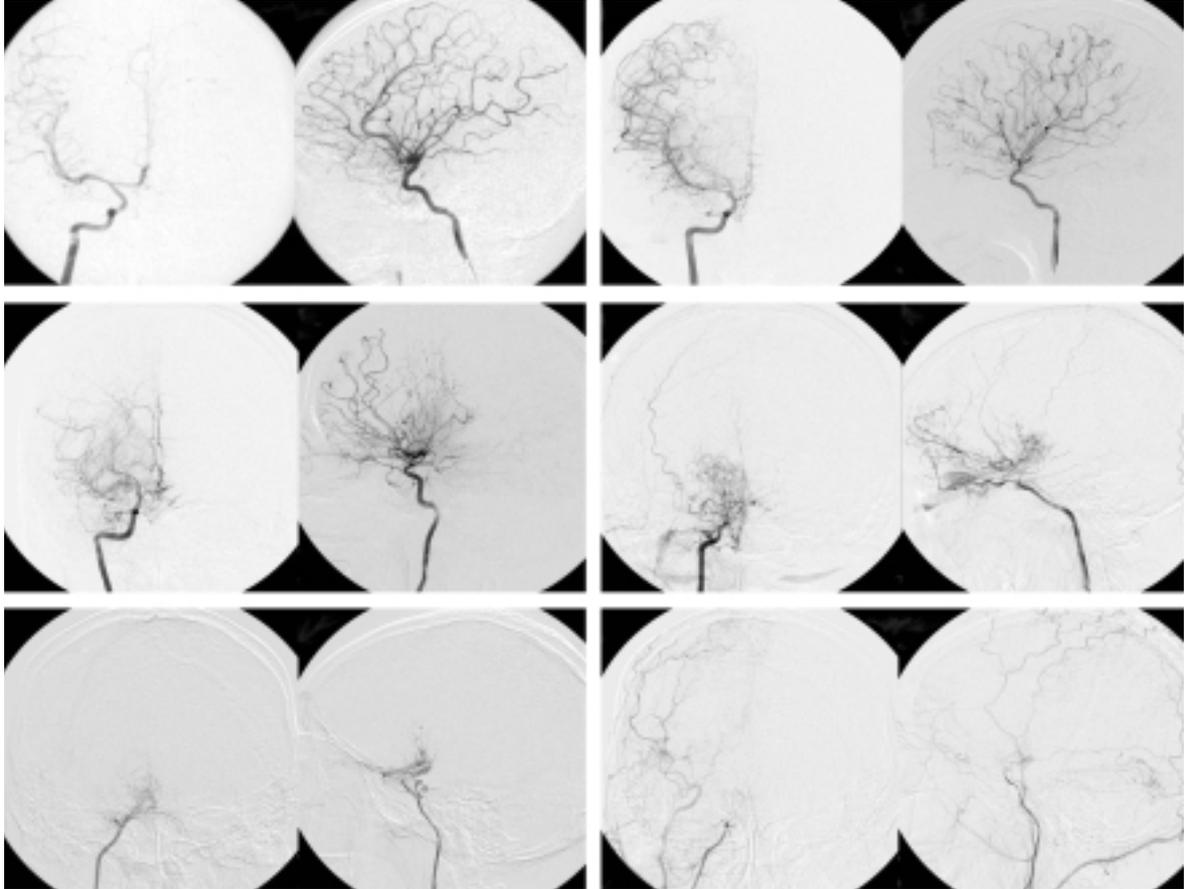
(unilateral moyamoya disease, quasi - moyamoya, akin - moyamoya, moyamoya syndrome, moyamoya phenomena, moyamoya - like vasculopathy)

PET PET 가 SPECT(single photon emission computed tomography)

(quasi - moyamoya disease) (rCBF) acetazolamide (perfusion reserve) (5).

(rCBF)가





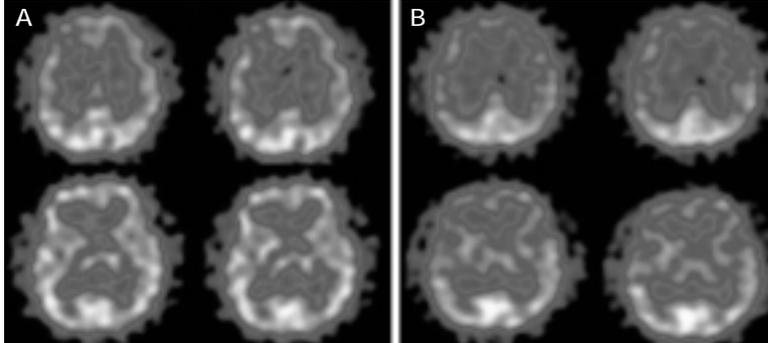
4. Suzuki stage

가 , 100,000
 3.16 0.35 , 1.8
 (7). 5 가 , 30~49
 . 8~10%
 가 . 가 , 10
 (juvenile type)가 50% , (adult
 type)

1.

가 .

44%



5. (A) SPECT (B) (acetazolamide)

(cerebral revascularization surgery) 가 (13~16).

2.4%

가

16.4% (8).

(transient ischemic attack, TIA)

가

(4, 5),

(9) 가

26%

11% 10 , 7% 24

, 3% (9). 5 가 (10)

(11, 12).

2.

가 ,

1) (Direct Revascularization) (superficial temporal artery) (middle cerebral artery) (anastomosis) (6). 가

(7).

가

(17,

18)

(19, 20),

가

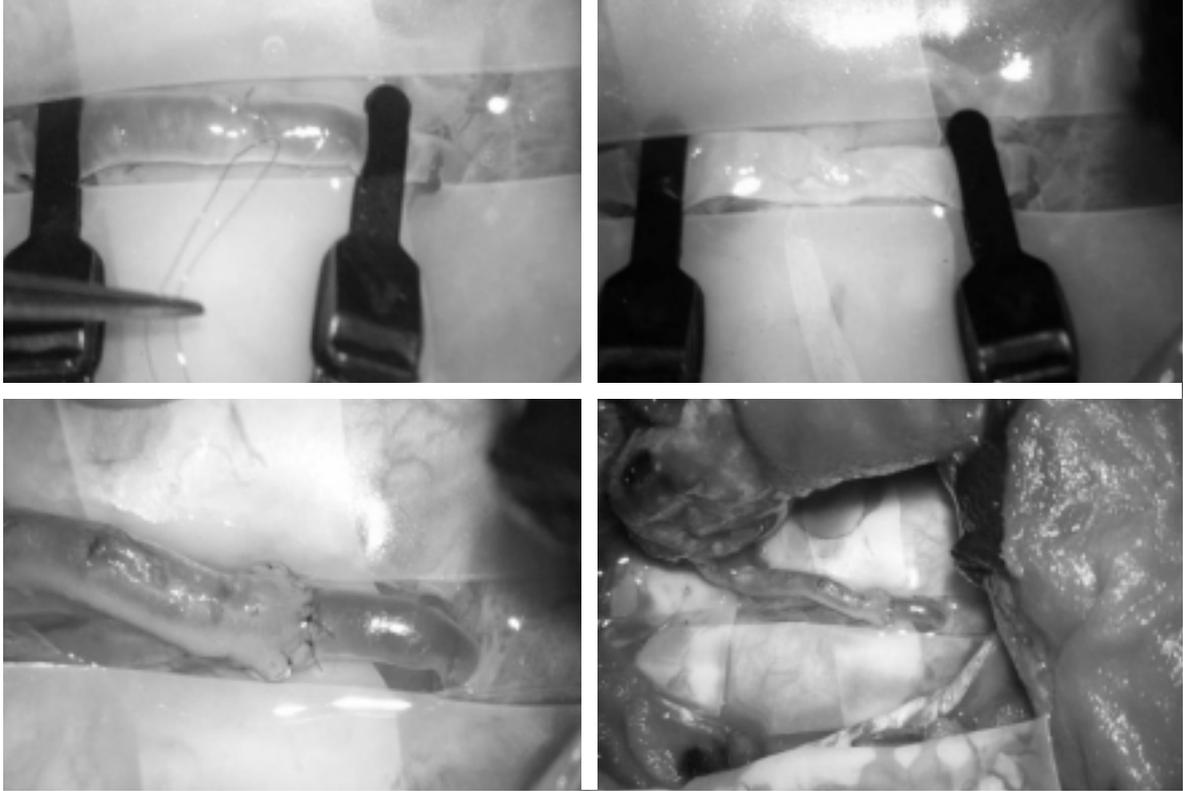
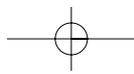
가

2) (Indirect Revascularization)

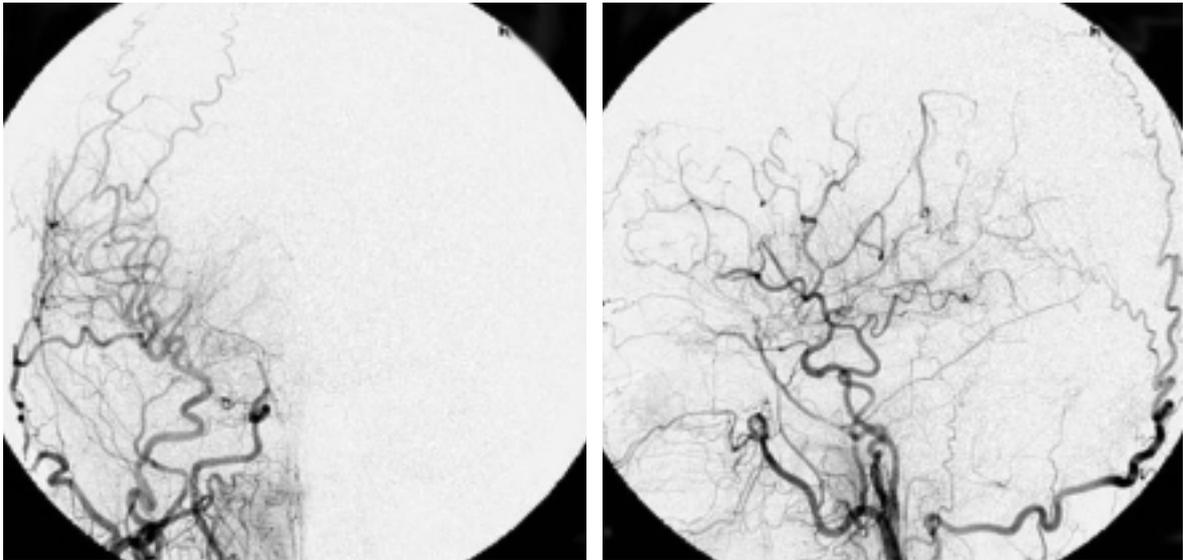
가

(galea aponeurotica), (angiogenesis or neovascularization) (,

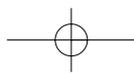
(syngangiosis)

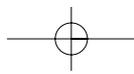


6. (STA - MCA anastomosis)

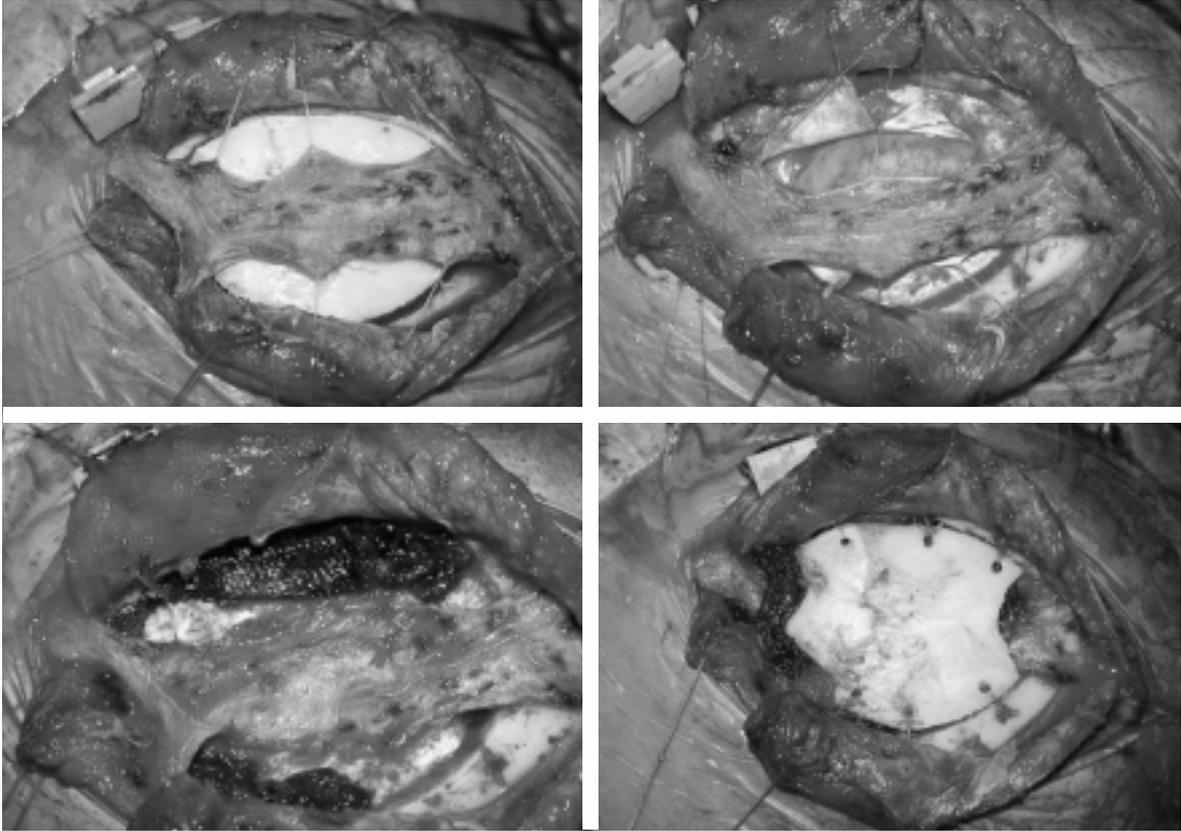


7. 7

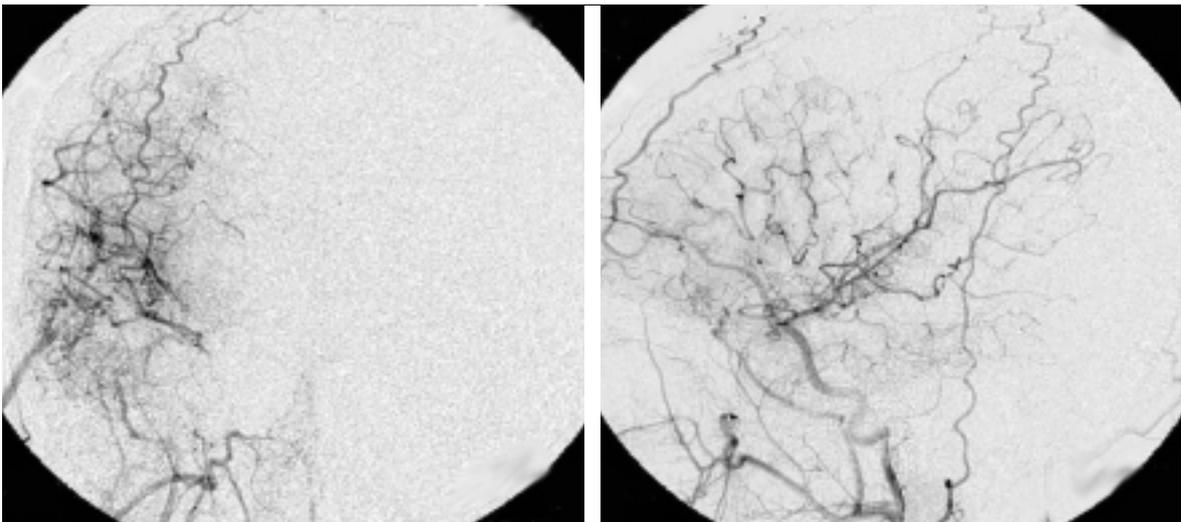




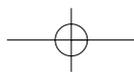
Continuing Education Column

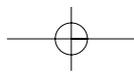


8. (EDAS=encephalo - duro - arterio - syngangiosis)



9. 6





(9), 가 3~5%
(21). ,
가 (17), 30~40 (15, 23),

3. 가 .
가 가 .
가 가 .
가 (14,
21, 22).

(21):

(21, 23), 가

가

(19, 21).

가

(7,

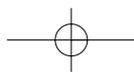
21).

(Japan

Adult Moyamoya Trial)가

4.

1. Takeuchi K, Shimizu K. Hypoplasia of the bilateral internal carotid arteries. No To Shinkei 1957; 9: 37 - 43
2. Nishimoto A, Takeuchi S. Abnormal cerebrovascular network related to the internal carotid arteries. J Neurosurg 1968; 29: 255 - 60
3. Suzuki J, Kowada M, Asahi M. Diseases showing the "fibrille" like vessels at the base of brain, particularly found in Japan. No To Shinkei 1965; 17: 767 - 76
4. Suzuki J, ed. Moyamoya disease. Tokyo: Springer - Verlag, 1986: 1 - 52
5. Suzuki J, Takaku A. Cerebrovascular "moyamoya" disease. Disease showing abnormal net - like vessels in base of brain. Arch Neurol 1969; 20: 288 - 99
6. Fukui M, Members of the Research Committee on Spontaneous Occlusion the Circle of Willis. Guidelines for the diagnosis and treatment of spontaneous occlusion of the circle of Willis(" moyamoya disease"). Clin Neurol Neurosurg 1997;



Continuing Education Column

- 99(Suppl 2): 238 - 40
7. Wakai K, Tamakoshi A, Ikezaki K, Fukui M, Kawamura T, Ohno Y, et al. Epidemiological features of moyamoya disease in Japan: findings from a nationwide survey. *Clin Neurol neurosurg* 1997; 99(Suppl 2): 1 - 5
 8. Yonekawa Y, Taub E. Moyamoya disease: status 1998. *Neurologist* 1999; 5: 13 - 23
 9. Kurokawa T, Tomita S, Ueda K, Narazaki O, Hanai T, Kitamura K, et al. Prognosis of occlusive disease of the circle of Willis (moyamoya disease) in children. *Pediatr Neurol* 1985; 1: 274 - 7
 10. Fukuyama Y, Umezu R. Clinical and cerebral angiographic evolutions of idiopathic progressive occlusive disease of the circle of Willis(" moyamoya disease") in children. *Brain Dev* 1985; 7: 21 - 37
 11. Ishii R, Takeuchi S, Ibayashi K, tanaka R. Intelligence in children with moyamoy disease: evaluation after surgical treatment with special reference to changes in cerebral blood flow. *Stroke* 1984; 15: 873 - 7
 12. Matsushima Y, Aoyagi M, Nariai T, Takada Y, Hirakawa K. Long - term intelligence outcome of post - encephalo - duro - arterio - synangiosis in childhood moyamoya patients. *Clin Neurol Neurosurg* 1997; 99(Suppl 2): 147 - 50
 13. Golby AJ, Marks MP, Thompson RC, Steinberg GK. Direct and combined revascularization in pediatric moyamoya disease. *Neurosurgery* 1999; 45: 50 - 60
 14. Houkin K, Kamiyama H, Takahashi A, Kuroda S, Abe H. Combined revascularization surgery for childhood moyamoya disease. STA - MCA and encephalo - duro - arterio - myo - synangiosis. *Childs Nerv Syst* 1997; 13: 24 - 9
 15. Karasawa J, Touho H, Ohnishi H, Miyamoto S, Kikuchi H. Long - term follow - up study after extracranial - intracranial bypass surgery for anterior circulation ischemia in childhood moyamoya disease. *J Neurosurg* 1992; 77: 84 - 9
 16. Matsushima T, Inoue t, Suzuki SO, Fujii K, Fukui M, Hasuo K. Surgical treatment of moyamoya disease in pediatric patients' comparison between the results of indirect and direct revascularization procedures. *Neurosurgery* 1992; 31: 401 - 5
 17. Houkin K, Kuroda S, Ishikawa T, Abe H. Neovascularization (angiogenesis) after revasculararization in moyamoya disease. Which technique is most useful for moyamoya disease? *Acta Neurochir (Wien)* 2000; 142: 269 - 76
 18. Mizoi K, Kayama T, Yoshimoto T, Nagamine Y. Indirect revascularization for moyamoya disease: is there a beneficial effect for adult patients? *Surg Neurol* 1996; 45: 541 - 9
 19. Okada Y, Shima T, Nishida M, Yamane K, Yamada T, Yamana-ka C. Effectiveness of superficial temporal artery - middle cerebral artery anastomosis in adult moyamoya disease: cerebral hemodynamics and clinical course in ischemic and hemorrhagic varieties. *Stroke* 1998; 29: 625 - 30
 20. Watanabe H, Ohta S, Oka Y, Kumon Y, Sakaki S, Tanada S, et al. Changes in cortical CBF and vascular response after vascular reconstruction in patients with adult onset moyamoya disease. *Acta Neurochir (Wien)* 1996; 138: 1211 - 7
 21. Houkin K, Kamiyama H, Abe H, Takahashi A, Kuroda S. Surgical therapy for adult moyamoya disease. Can surgical revascularization prevent the recurrence of intracerebral hemorrhage? *Stroke* 1996; 27: 1342 - 6
 22. Houkin K, Ishikawa T, Yoshimoto T, Abe H. Direct and indirect revascularization for moyamoya disease. Surgical techniques and peri - operative complications. *Clin Neurol Neurosurg* 1997; 99(Suppl 2): 142 - 5
 23. Ishikawa T, Houkin K, Kamiyama H, Abe H. Effects of surgical revascularization on outcome of patients with pediatric moyamoya disease. *Stroke* 1998; 28: 1170 - 3