대한뇌혈관내수술학회 | 대한뇌혈관외과학회

SKEN-KSCVS
합동 연수강좌

일 시 | **2019년 9월 7일(토)**
장 소 | 분당서울대병원 헬스케어 혁신파크 4층 미래홀
주 최 | 대한뇌혈관내수술학회, 대한뇌혈관외과학회
주 관 | 대한신경외과학연구재단
평 점 | 6점

대한뇌혈관내수술학회
SKEN
Society of Korean Endovascular Neurosurgeons

Korean Society
of Cerebrovascular Surgeons
대한뇌혈관외과학회

2019 대한뇌혈관내수술학회 임원진

명예회장

직 위	성 명	소 속
명예회장	백민우	인봉의료재단 뉴고려병원

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2019 대한뇌혈관내수술학회 임원진

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인사말

대한뇌혈관내수술학회와 대한뇌혈관외과학회 회원 여러분 안녕하십니까!

우리 모두를 힘들게 했던 한여름 폭염도 점차 지나가고 있고 모든 것이 풍성해지는 가을이 어김없이 다가오고 있습니다.

먼저 이번 2019년 대한뇌혈관내수술학회와 대한뇌혈관외과학회 합동연수강좌를 함께 준비해주신 대한뇌혈관외과학회 김종수 회장님과 양 학회 이사진 여러분의 노고에 깊이 감사드립니다. 뇌혈관질환을 전문적으로 다루는 대한뇌혈관외과학회와 대한뇌혈관내수술학회가 서로의 경험을 공유하고 새로운 술기와 지식을 습득하는 것은 양 학회 회원들의 질적 향상뿐 아니라 아직까지도 국내에서 높은 사망률을 보이는 뇌혈관질환 관련 환자들에게 최선의 진료를 제공할 수 있는 필수적인 과정이라 판단됩니다. 이번 합동연수강좌에서는 모야모야병과 두개강내 뇌혈관박리질환에 대해 심도 있는 강의를 준비했고 뇌졸중분야에서 DTI의 임상적 적용에 관해 전문가를 모시고 토론하는 시간을 가질 예정입니다. 아무쪼록 알차고 의미 있는 연수강좌가 될 수 있기를 기대하며 양 학회 회원 간의 친교도 돈독히 할 수 있는 뜻 깊은 시간이 되기를 기대합니다.

다시 한번 이번 2019 대한뇌혈관내수술학회–대한뇌혈관외과학회 합동연수강좌를 위해 수고해주신 모든 여러분들께 감사드립니다.

대한뇌혈관내수술학회 회장

고 준 석

인사말

친애하는 대한뇌혈관내수술학회 및 대한뇌혈관외과학회 회원 여러분!

수확을 앞둔 결실의 이 좋은 계절에 양 학회가 합동 연수강좌를 개최합니다.
올해는 대한뇌혈관내수술학회가 주최가 되어 모야모야병 및 IC dissection을 주제로 고준석 회장님과 여러 임원들이 수고해 주셨고 알찬 연수교육이 되리라 기대합니다.
본디 대한뇌혈관외과학회에서 출발한 회원님들이 뇌혈관내수술의 새로운 지평을 열고자 대한뇌혈관내수술학회를 만드시고 발전시켜 오신 데 대해 축하의 인사를 보냅니다.
최근의 양 학회 회원들은 두 학회 모두에서 활동하시고 계신 것으로 압니다. 향후 양 학회의 발전적 통합이 이루어져서 신경외과 뇌혈관 분야 전문의의 위상이 더욱 높아지고 협력과 연대를 바탕으로 세계를 선도하는 학회로의 발전을 꿈꾸어 봅니다.
양 학회 회원 모두의 정진과 발전을 기원합니다.

대한뇌혈관외과학회 회장
김 종 수

프로그램

대한뇌혈관내수술학회 | 대한뇌혈관외과학회

SKEN-KSCVS 합동 연수강좌

일 시 | 2019년 9월 7일(토)

장 소 | 분당서울대병원 헬스케어 혁신파크 4층 미래홀

09:15-09:25	개회사	고준석 (대한뇌혈관내수술학회 회장) 김종수 (대한뇌혈관외과학회 회장)	
09:25-09:30	축사	오창완 (대한신경외과학회 이사장)	
09:30-10:45	**Session I. Moyamoya Disease: General**	좌장: 강현승 (서울대), 안재성 (울산대)	
	Epidemiology and clinical course of Moyamoya disease	김재훈 (을지대)	16
	Genes in Moyamoya disease: Updates information of the function of RNF213 and novel candidate gene for Moyamoya disease	심규원 (연세대)	23
	Diagnostic image of Moyamoya disease: Radiologic image & differential diagnosis of Moyamoya disease	김용배 (연세대)	33
10:45-11:00	**Coffee Break**		
11:00-12:15	**Session II. Moyamoya Disease: Treatment**	좌장: 성재훈 (가톨릭대), 고현송 (충남대)	
	Bypass strategies for Moyamoya disease: Risk and incidence of complication & avoidance of complication from revascularization surgery	연제영 (성균관대)	36
	Surgical role in hemorrhagic Moyamoya disease	김정은 (서울대)	38
	Endovascular treatment for targeted embolization in Moyamoya disease with hemorrhagic manifestation	반승필 (서울대)	48
12:15-13:10	**Lunch**		
13:10-14:00	**Special Lecture**	좌장: 고준석 (경희대), 김종수 (성균관대)	
	Clinical application of DTI for stroke in the viewpoint of neurosurgery	장성호 (영남대 재활의학과)	62
14:00-15:15	**Session III. Intracranial Dissection: General**	좌장: 윤석만 (순천향대), 김태선 (전남대)	
	Epidemiology and pathophysiology of intracranial dissection	권오기 (서울대)	70
	Diagnostic image of intracranial dissection	류창우 (경희대 영상의학과)	71
	Natural course and medical treatment for intracranial dissection	김대원 (원광대)	75
15:15-15:30	**Coffee Break**		
15:30-17:00	**Session IV. Intracranial Dissection: Treatment**	좌장: 신용삼 (가톨릭대), 정영균 (인제대)	
	Surgical treatment for intracranial dissection	박재찬 (경북대)	74
	Endovascular treatment for intracranial dissection	백승국 (부산대 영상의학과)	79
	Complicated cases and panel discussion	주성필 (전남대)	83
		김성림 (가톨릭대)	86
		이재일 (부산대)	87
		박중철 (울산대)	88
17:00	폐회사	김종수 (대한뇌혈관외과학회 회장) 고준석 (대한뇌혈관내수술학회 회장)	

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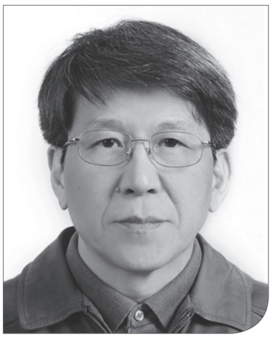

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현재 부산대 의학전문대학원 부교수

Session I. Moyamoya Disease: General

좌장: 강현승(서울대), 안재성(울산대)

1. Epidemiology and clinical course of Moyamoya disease
김재훈 (을지대)

2. Genes in Moyamoya disease: Updates information of the function of RNF213 and novel candidate gene for Moyamoya disease
심규원 (연세대)

3. Diagnostic image of Moyamoya disease: Radiologic image & differential diagnosis of Moyamoya disease
김용배 (연세대)

Epidemiology and clinical course of Moyamoya disease

김재훈

을지대

A. Definition

1. Moyamoya disease (MMD)

- Progressive stenosis and occlusion of the distal part of ICA
- Formation of a fine vascular network (moyamoya vessels)
- No etiology
- Bilateral lesions

2. Probable MMD or unilateral MMD

- Unilateral lesion

3. Quasi-MMD or MM syndrome

- Atherosclerosis, autoimmune disease, meningitis, brain neoplasm, Down's syndrome, neurofibromatosis, head trauma, cranial irradiation, sickle cell disease, hyperthyroidism

B. Epidemiology

1. Incidence (I) and prevalence (P)

- Korea : I = 1.0 (2008) ~ 4.3 (2013) /105-year
  P = 6.3 (2005) ~ 18.1 (2013) /105
- Japan : I = 0.35 (1994) ~ 1.36 (2006) /105-year
  P = 3.16 (1994) ~ 10.5 (2002-2006) /105
- China : I = 0.43 (2000-2007) ~ /105 –year
  P = 3.92 (2000-2007) ~ /105
- Taiwan : I = 0.048 (1978) ~ 0.2 (2011) /105-year

P = 1.61(2011) /105

- USA : I = 0.05 (1973-1997) ~ 0.17 (1987-1996) /105-year
- Europe : I = 0.03 (1996) ~ 0.047 (1994-2015) /105-year

2. Gender differences (Female to Male ratio)

- Korea : 1.4 (2010) ~ 1.9 (2004-2008)
- Japan : 1.8 (1994) ~ 2.2 (2002-2006)
- China : 0.9 (2002-2010) ~ 1.1 (2000-2007)
- Taiwan : 1.1 (2000-2005) ~ 1.7 (2006-2011)
- USA : 2.5 ~ 8
- Europe : 2.1 (1994-2015) ~ 4.2 (1996-2007)
- Explanations: Difference in coagulation and fibrinolytic pathways, the effect of estrogen on the endothelium and surrounding connective tissue, X-linked genetic differences, methylation patterns

3. Age distribution (years)

- Korea : 2 peaks (5-15 vs. 45-54)
- Japan : 2 peaks (5-9 vs. 45-49)
- China : 2 peaks (5-9 vs. 35-45)
- Taiwan : 2 peaks (5-14 vs. 40-44)
- USA : 2 peaks (5-9 vs. 55-59)
- Europe : 2 peaks (0-9 vs. 30-39)

4. Familial MMD

- Autosomal dominant inheritance with incomplete penetration
- Familial occurrence: 10-15% (East Asian), 2% (USA), 1.3-7.1% (Europe)
- Early onset of symptom
- Higher Female to male ratio

5. Explanations for the discrepancy of epidemiology

- Different survey methods (different inclusion criteria/ population based or in-hospital based)
- Different genetic or environmental factor

6. Trends

- Increase of both the incidence and prevalence because of an increase in newly diagnosed MMD and survivors
- Change of epidemiologic features

C. Clinical course

1. TIA and ischemic stroke

- Cerebral ischemia in the territory of the ICA, particularly in the frontal lobe
- Focal neurological deficit: hemiparesis, speech or sensory disturbances
- Atypical symptoms: syncope, visual symptoms, involuntary movements, intellectual or cognitive impairment
- Hyperventilation-induced ischemic attack in pediatric patients

2. Hemorrhage

- Intraventricular (IV), intraparenchymal (IP), subarachnoid hemorrhage
- IVH occurs more commonly than in hypertensive IPH
- Rupture of dilated, fragile MM vessels: persistent hemodynamic stress of the MM vessels (basal ganglia, thalamus, or periventricular region)
- Rupture of saccular aneurysms: hemodynamic stress induces the formation of a saccular aneurysm
- Rupture of dilated pial collateral arterioles

3. Headache

- Frontal or migraine-like headache
- Lower the threshold for migraine or increase spreading cortical depression
- Dilated transdural collaterals stimulate dural nociceptors

4. Seizure

- 20-30% of MMD patients
- Predictors for epilepsy: lower mRS score, diffuse brain atrophy, early seizure

5. Involuntary movement

- Usually in children (4% of pediatric patients)
- Cerebral hypoperfusion in the basal ganglia and in the relevant cerebral cortex
- dilated MM-associated collaterals in BG

6. Intellectual or cognitive impairment

- Intellectual impairment due to frontal ischemia in pediatric patients
- Cognitive dysfunction (delayed recall, visual space & executive function) in adults due to frontal, medial temporal, hippocampal ischemia
- Ongoing COSMO-JAPAN study

D. Interesting issues

1. Natural course

1) Kuroda (2005)
- MMD progression: 15/63 (23.8%)
- Mean time to progression from diagnosis: 60 months (1.5-8 years)
- Risk factor: female

2) Kuroda (2007)
- MMD progression: related to silent infarction or ischemic events
- Annual stroke rate = 3.2% (mean FU 43.7 months)

3) Gross (2013)
- 42 MMD/S patients, mean FU 2.9 years
- Annual stroke rate = 13.3%, hemorrhage rate = 1.7%
- Risk factors for future stroke: female, recent stroke (within 3 years)

4) Cho (2015)
- 241 adults with stable hemodynamics, (mean FU 82.5 months)
- Type: ischemia (59.8%), hemorrhage (25.7%), asymptomatic (14.5%)
- Annual stroke rate = 4.5% (ischemic 3%, hemorrhagic 4.3%)
- Annual stroke rate in asymptomatic patients = 3.4% (ischemic 0.8%, hemorrhagic 2.5%)
- Worst clinical outcomes: hemorrhagic group

5) Kim (2017)
- 176 patients, mean FU 83 months
- Overall annual recurrent hemorrhage rate = 3.4%/person during 5 years

2. Unilateral MMD

1) Smith (2008)
- 10/33 patients progressed to bilateral disease over 5 years
- disease progression risk: contralateral abnormalities on initial imaging, congenital cardiac anomalies, previous cranial irradiation, Asian ethnic origin, familial MMD

2) Hayashi (2013)
- Incidence = 0.23/105-year, prevalence = 0.66/105
- Ratio of unilateral to all MMD: 10.6%

3. MMD-related aneurysm (3.4~14.8%)

1) Yeon (2011)
   - Prevalence = 9/249 (3.6%)
   - Posterior location: 54%
   - Increasing tendency in patients over 50 years

2) Zhang (2015)
   - Pathogenesis: hemodynamic stress, pathological vessel architecture
   - Type: major artery, peripheral artery (ant. & post. Choroidal artery), moyamoya vessel (lenticulostriate, thalamo-perforating artery), meningeal artery, anastomosis site

3) Kim (2018)
   - Prevalence = 11/132 (8.3%)
   - Rupture rate = 7/11 (63.6%)

4. Asymptomatic MMD

1) Ikeda (2006)
   - 8/11402 healthy (0.07%)
   - Men 0.05%, Women 0.1%
   - Mean age: men 54.8 years, women 53.3 years
   - Estimated prevalence 50.7/105
   - Family history: 62.5%

2) Kuroda (2007)
   - 40 asymptomatic patients (13-67 years)
   - Silent infarction: 20%, Disturbed cerebral hemodynamics: 40%
   - 7/34 conservatively treated patients: TIA (3), intracranial bleeding (3), ischemic stroke (1) 43.7 months FU
   - Annual risk for any stroke: 3.2%
   - Asymptomatic MMD: not a silent disorder and potentially cause ischemic or hemorrhagic stroke.

3) Jo (2014)
   - 40 (74 hemispheres) asymptomatic adult patients
   - 32 months FU: TIA 3/40
   - Angiographic 24 months FU: 3 progression, 3 new hemodynamic abnormality
   - Predictors of symptomatic progression: decreased vascular reserve capacity, smoking

4) Yang (2014)
- 42 (75 hemispheres) asymptomatic adult patients (37 months FU)
- 4 Hemisphere (5.3%) symptomatic progression (3 hemorrhage, 1 TIA)
- 9 Hemisphere (12%) angiographic progression
- Predictor of progression: decreased initial cerebrovascular reserve capacity

5) Ongoing AMORE study

5. Quasi-MMD or MM syndrome

1) Hayashi (2014)
- 109 patients
- F:M = 1.57, mean age = 30.6 years
- Bimodal peaks, familial (7%)
- Cause: atherosclerosis, Down syndrome, von Recklinghausen disease
- Unilateral lesion, mild moyamoya vessels, less frequent intracranial hemorrhage

2) Zhao (2017)
- 64/693 patients (9.2%)
- Mean age: 31.5 years, unimodal age distribution
- Cause: atherosclerosis, hyperthyroidism
- Annual risk of stroke = 19.4%/patient

E. Future perspectives

- Epidemiology: need to perform a population-based study, especially in Westerns
- Natural history of MMD (symptomatic, asymptomatic, disease progression): multicenter prospective observational trial

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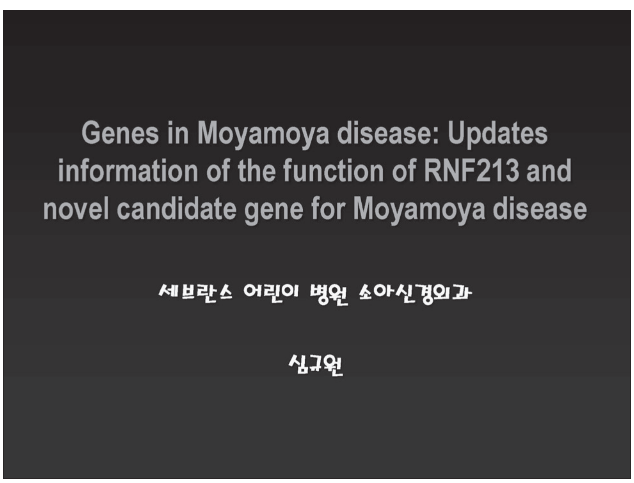
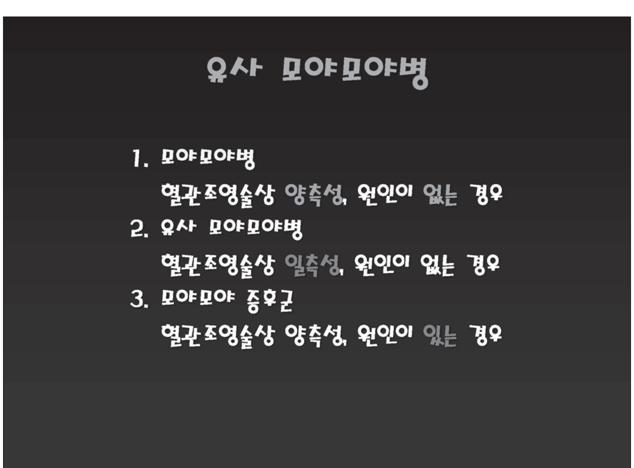
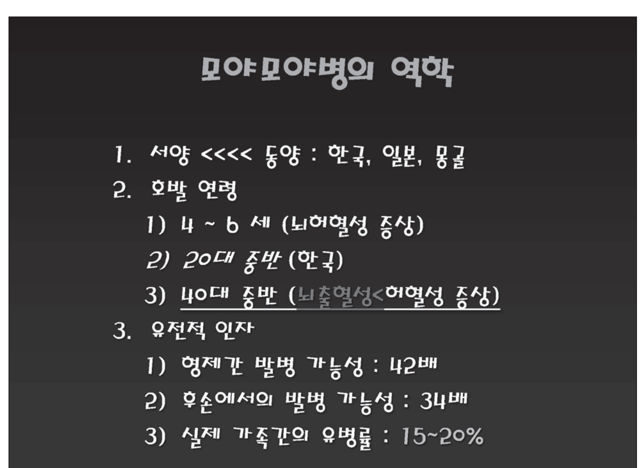
Genes in Moyamoya disease: Updates information of the function of RNF213 and novel candidate gene for Moyamoya disease

심 규 원

연 세 대







Epidemiology

Japan

	Incidence/Prevalence	Patients (*N*)	Female-male	Familial (%)	Age at onset
1995	0.35/3.16	5,227	1.8	10	5 yr 30-49 yr
2006	0.94/10.5	10,812	2.18	15.4	45-49 yr 5-9 yr

2006; 2008 JNNP Baba + 1997 CNN Wakai

Epidemiology

Japan

Table 1 Epidemiological features of moyamoya disease

	Present study	Previous study—1997[2]
Number of cases	267	1176
Detection rate (per 100 000)	0.94	0.35
Prevalence (per 100 000)	10.5	3.16
Sex ratio (male:female)	1:2.2	1:1.8
Patients younger than 10 years old at onset (%)	15.1%	47.8%
Pattern of age distribution	2 peaks	2 peaks
Highest peak in age distribution (in years)	45–49	10–14
Second peak in age distribution (in years)	5–9	40–49
Patients with family history	15.4%	10.0%

종전의 결과에 비해 몇 가지의 차이를 보인다.
Detection rate가 높아짐.
10세 이하는 15.1%. 10세 이전에 증세가 나타나는 비율이 줄었음.
가장 많이 진단되는 나이는 45-49세다. 나이가 조금 달라짐.
여 : 남 = 2.18 : 1. 성인이세 여성 환자의 비율이 두드러지는 특징을 보인다.

2008 JNNP Baba T., Houkin K, et al

Epidemiology

Japan

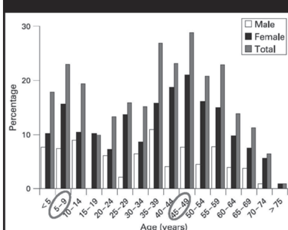


Figure 2 Onset age distribution of moyamoya disease.

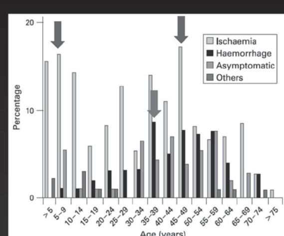


Figure 3 Age distribution of disease pattern at onset.

허혈형 57.4%, two peak 45-49, 5-9 ( )
출혈형 21%, 35-39세에 one peak ( )
무증상 17.8%, 5-8, adult

Epidemiology

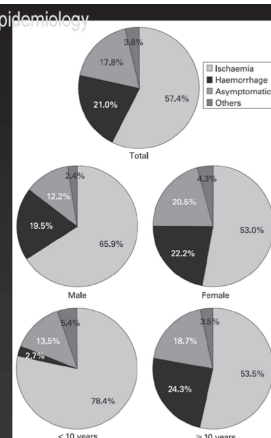


Figure 4 Disease patterns of moyamoya disease at onset.

Ischemia; male : female= 65.9% : 53%

Female; hemorrhage; 22.2%, asymptomatic; 20.5%
Male; hemorrhage;19.5%, asymptomatic;12.2%

Ischemia; 78.4% <10yrs, 53.5% > 10 yrs.
Hemorrhage; 2.7% <10yrs, 24.3% > 10 yrs.
Familial MMD; 37.8% < 10yrs, 12.2% > 10 yrs

종전의 자료는 주로 대학 병원에서 받은 것들이지만 이번 연구는 중 종합병원 및 다양한 형태의 지역병원에서 자료를 받았다.
따라서 이전에 자료에서는 누락되었던 성인 환자의 자료가 많이 포함되었을 것이다.

18%의 무증상 환자들이 특이한데, 이는 일본의 brain check-up system으로 인한 특이점이라 할 수 있다.

Epidemiology

2000 Korea

Table 1. *General Demographic Data of the Patients*

Characteristics	All patients	Adults (=>16)	Children (<16)	p-value**
Number (%)	334	213 (63.8%)	121 (36.2%)	
Age, mean ± SD, year	26.7 ± 17.1	37.1 ± 12.1	8.2 ± 3.6	
Median, year	26.5	36.1	3.6	
Sex, % of male	40.7	39.9	42.1	NS
Mode of presentation				<0.0001
Haemorrhage, %	43.1	62.4	9.1	
Ischaemia, %	38.3	25.4	61.2	
Seizure, %	10.8	3.8	23.1	
Presenting symptoms				
Decreased level of consciousness, %	38.0	54.5	9.1	<0.0001
Weakness, %	59	45.5	83.5	<0.0001
Suzuki grade, mean ± SD	3.4 ± 1.1	3.4 ± 1.2	3.4 ± 0.9	NS
Favorable onset (KPS => 70), %	73.1	57.3	80.2	<0.0001
Mode of treatment				
Bypass surgeries, %	38.1	24.4	62.0	<0.0001
(Bilateral bypass surgery (%))*	(53.5)	(46.2)	(58.7)	NS
Outcome				
Mortality, %	3.3	4.7	0.8	<0.0001
Favorable outcome (KPS => 70), %	73.1	70.4	77.7	0.09

* Percentage of bilateral bypass surgery in cases performed bypass operations.
** Comparison between adult and child patients.
NS Statistically not significant.

2000 Acta

Epidemiology

2000 Korea

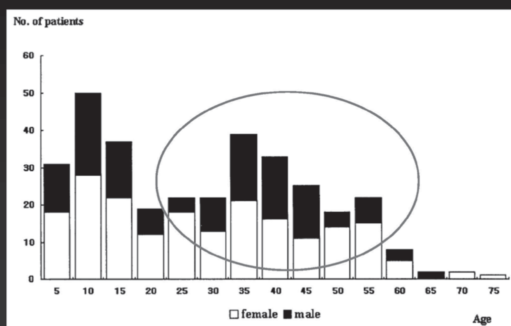
Table 2. *Comparisons of Clinical Characteristics Between Haemorrhage and Ischaemia Group*

Characteristics	Haemorrhage	Ischaemia	p-value
Number	144	126	
Age, mean ± SD, year	36.9 ± 13.1	19.2 ± 16.0	<0.0001
Median, year	37	12	
% of adults	92.4	42.2	<0.0001
Sex, % of male patients	35.4	44.5	NS
Presenting symptoms			
Decreased level of consciousness, %	63.2	14.8	<0.0001
Weakness, %	39.6	80.5	<0.0001
Suzuki grade, mean ± SD	3.4 ± 1.4	3.5 ± 0.9	NS
Favorable onset (KPS => 70), %	52.8	78.9	<0.0001
Mode of treatment			
Bypass surgeries, %	25.7	53.9	<0.0001
(Bilateral bypass, %)*	(43.2)	(53.6)	NS
Outcome			
Mortality, %	5.8	2.3	<0.0001
Favorable outcome (KPS => 70), %	67.4	77.3	NS

* Percentage of bilateral bypass surgery in cases performed bypass operations.
NS Statistically not significant.

Epidemiology

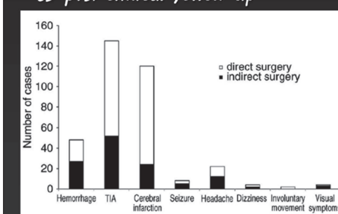
2000 Korea

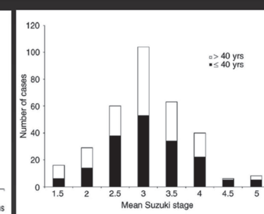


Epidemiology

2012 Korea

348 pts (M : F = 128 : 220), Age 39 years (18-68 years)
325 surgical treatment for 283 pts
  216 direct + indirect for 192 pts
  109 direct for 91 pts
65 pts; clinical follow-up





2012 Neuro Med Chir JE Kim et al

원인 : ????????????

1) 나병, 결핵 등의 감염
2) 섬유아세포 증식 인자
3) 면역학적 질환
4) 혈전
5) 교감신경 이상
6) 유전적 이상 : Phakomatosis, fibromuscular dysplasia, collagen disease, Down syndrome, MMP9, RNF213?
7) 혈관의 내막 손상 - platelet derived growth factor (PDGF) – 내경동맥 협착
8) 혈관 내피세포 이상?

병인론

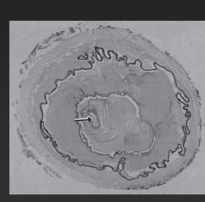
1. 혈소판 유발 인자의 혈관 내피 치유 과정의 이상
2. 섬유 아세포 성장인자의 혈관 손상치유 과정의 이상

-> 혈관 내피 세포의 과다 증식으로 점진적인 혈관 막힘 유발

조직학적 이상 소견

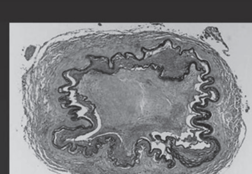
일차 병변 (뇌혈관벽의 내피층)

1) 섬유소의 침착으로 내피 두꺼워짐
2) 내측 탄력층의 접힘과 구부러짐, 늘어짐
3) 염증 소견은 거의 없다.



이차 측부순환

1) 내측 탄력층의 파괴
2) 내피의 섬유화
3) 미세 동맥류
4) 부분 파열



Smooth-muscle progenitor cells isolated from patients with moyamoya disease: novel experimental cell model

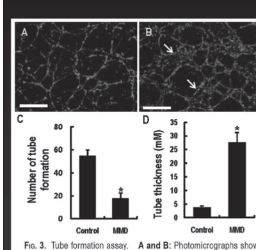


FIG. 3. Tube formation assay. **A and B:** Photomicrographs showing immunofluorescence staining of SPCs. Compared with SPCs obtained in a healthy volunteer (**A**), SPCs obtained in a patient with MMD made rather irregularly arranged tubules of varying sizes (**B**). In some areas, thickened tubules were noted *(arrows)*. Bars = 500 μm. **C and D:** Bar graphs showing that SPCs obtained in patients with MMD formed thickened tubules of fewer numbers compared with controls (SPCs were obtained in 3 patients and 3 control volunteers). *Asterisks* = $p < 0.01$.

TABLE 2: Gene ontology terms in upregulated genes in SPCs obtained in patients with MMD

Term	p Value
GO:0009719; response to endogenous stimulus	0.016
GO:0003002; regionalization	0.017
GO:0042493; response to drug	0.023
GO:0009952; anterior/posterior pattern formation	0.033
GO:0048013; ephrin receptor signaling pathway	0.035
GO:0009725; response to hormone stimulus	0.037
GO:0010243; response to organic nitrogen	0.040
GO:0043434; response to peptide hormone stimulus	0.042
GO:0007389; pattern specification process	0.045
GO:0000002; mitochondrial genome maintenance	0.054
GO:0030031; cell projection assembly	0.065
GO:0030182; neuron differentiation	0.069
GO:0010033; response to organic substance	0.069
GO:0008202; steroid metabolic process	0.081
GO:0001822; kidney development	0.084
GO:0030324; lung development	0.089
GO:0030323; respiratory tube development	0.093
GO:0035295; tube development	0.099
GO:0042127; regulation of cell proliferation	0.099

TABLE 3: Gene ontology terms in downregulated genes in SPCs obtained in patients with MMD

Term	p Value
GO:0007155; cell adhesion	0.0000385
GO:0022610; biological adhesion	0.0000392
GO:0010033; response to organic substance	0.0000565
GO:0030334; regulation of cell migration	0.000122
GO:0045087; innate immune response	0.000214
GO:0040012; regulation of locomotion	0.000291
GO:0051270; regulation of cell motion	0.000302
GO:0007167; enzyme-linked receptor protein signaling pathway	0.000868
GO:0051605; protein maturation by peptide bond cleavage	0.000956
GO:0002684; positive regulation of immune system process	0.0012
GO:0003013; circulatory system process	0.0013
GO:0008015; blood circulation	0.0013
GO:0032535; regulation of cellular component size	0.0027
GO:0016485; protein processing	0.0031
GO:0051604; protein maturation	0.0044
GO:0006959; humoral immune response	0.0051
GO:0006956; complement activation	0.0059
GO:0002541; activation of plasma proteins involved in acute inflammatory response	0.0063
GO:0001944; vasculature development	0.0067
GO:0009611; response to wounding	0.0070
GO:0006955; immune response	0.0076
GO:0030335; positive regulation of cell migration	0.0078
GO:0007160; cell-matrix adhesion	0.0078
GO:0006952; defense response	0.0079
GO:0050778; positive regulation of immune response	0.0091
GO:0002253; activation of immune response	0.0095

J Neurosurg 120:415–425, 2014

TABLE 5: Literature review of putative pathogenetic DEGs in SPCs related to MMD*

Genes & Expression	Putative Pathogenetic Roles	Authors & Year
↑*ITGA3*	defective vascular morphogenesis	Ozerdem et al., 2001
↑*BAIAP2L1*	occlusion of internal carotid artery terminus	Koh et al., 2004; Nishimori et al., 1997
↑*CDH2*	SMC phenotype change & increasing mobilization	Tillet et al., 2005
↑*EPHA5*	defective vessel wall assembly	Kuijper et al., 2007; Zhang & Hughes, 2006
↑*MCAM*	vascular SMC proliferation	Okumura et al., 2004; Shih & Kurman, 1996
↑*GARNL4*	impairment of postnatal angiogenesis	Carmona et al., 2009; Nishimoto, 2000
↓*PLXNC1*	disruption of vascular tree formation & increased cellular migration & intimal hyperplasia	Walzer et al., 2005
↓*NEO1*	interruption of vessel wall maturation	Park et al., 2004
↓*ARHGAP28*	sustained activation of Rho GTPases & increased SMC migration & proliferation	Etienne-Manneville & Hall, 2002

* *ARHGAP28* = Rho GTPase-activating protein 28; *BAIAP2L1* = BAI1-associated protein 2–like 1; *CDH2* = N-cadherin; *EPHA5* = Eph receptor A5; *GARNL4* = GTPase-activating Rap/RanGAP domain–like 4; *ITGA3* = integrin α3; *NEO1* = neogenin homolog 1; *PLXNC1* = plexin C1; ↑ = upregulated; ↓ = downregulated.

J Neurosurg 120:415–425, 2014

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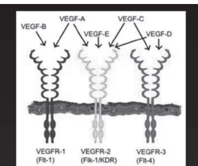
The Role of *VEGF* and *KDR* Polymorphisms in Moyamoya Disease and Collateral Revascularization

Table 7. Comparison of the *VEGF* and *KDR* genotype frequencies according to collateral vessel formation score.

Characteristics	Collateral grade A (n = 40)	Collateral grade B and C (n = 24)	AOR (95% CI)	P*
VEGF −2578C>A				
CC	20 (50.0)	13 (54.2)	1.000 (reference)	
CA	18 (45.0)	7 (29.2)	0.615 (0.200–1.893)	0.397
AA	2 (5.0)	4 (16.7)	3.060 (0.473–19.816)	0.241
Dominant (CC vs. CA+AA)			0.867 (0.313–2.408)	0.785
Recessive (CC+CA vs. AA)			3.757 (0.625–22.590)	0.148
VEGF −1154G>A				
GG	27 (67.5)	17 (70.8)	1.000 (reference)	
GA	12 (30.0)	6 (25.0)	0.841 (0.253–2.796)	0.777
AA	1 (2.5)	1 (4.2)	1.475 (0.082–26.410)	0.792
Dominant (GG vs. GA+AA)			0.926 (0.297–2.886)	0.895
Recessive (GG+GA vs. AA)			1.565 (0.090–27.244)	0.759
VEGF −634G>C				
GG	11 (27.5)	13 (54.2)	1.000 (reference)	
GC	22 (55.0)	7 (29.2)	0.252 (0.076–0.837)	0.024
CC	7 (17.5)	4 (16.7)	0.284 (0.045–1.780)	0.179
Dominant (GG vs. GC+CC)			0.286 (0.095–0.859)	0.026
Recessive (GG+GC vs. CC)			0.857 (0.215–3.412)	0.827
VEGF 936C>T				
CC	27 (67.5)	16 (66.7)	1.000 (reference)	
CT	13 (32.5)	8 (33.3)	1.069 (0.357–3.206)	0.905
TT	0 (0.0)	0 (0.0)	NA	NA
Dominant (CC vs. CT+TT)			1.069 (0.357–3.206)	0.905
Recessive (CC+CT vs. TT)			NA	NA
KDR −604T>C				
TT	17 (42.5)	11 (45.8)	1.000 (reference)	
TC	19 (47.5)	12 (50.0)	0.950 (0.325–2.778)	0.926
CC	4 (10.0)	1 (4.2)	0.370 (0.035–3.962)	0.411
Dominant (TT vs. TC+CC)			0.840 (0.298–2.367)	0.741
Recessive (TT+TC vs. CC)			0.346 (0.035–3.409)	0.363
KDR 1192G>A				
GG	30 (75.0)	16 (66.7)	1.000 (reference)	
GA	8 (20.0)	7 (29.2)	1.381 (0.401–4.758)	0.609
AA	2 (5.0)	1 (4.2)	0.843 (0.063–11.263)	0.897
Dominant (GG vs. GA+AA)			1.355 (0.420–4.375)	0.612
Recessive (GG+GA vs. AA)			0.664 (0.053–8.390)	0.752
KDR 1719T>A				
TT	10 (25.0)	7 (29.2)	1.000 (reference)	
TA	18 (45.0)	9 (37.5)	0.786 (0.219–2.824)	0.712
AA	12 (30.0)	8 (33.3)	0.886 (0.227–3.462)	0.861
Dominant (TT vs. TA+AA)			0.825 (0.259–2.626)	0.745
Recessive (TT+TA vs. AA)			1.195 (0.398–3.593)	0.751

*Adjusted by age and gender. NA; not applicable. AOR, adjusted odds ratio. CI, confidence interval.



- Endogenous VEGF production mediates compensatory revascularization during various physiological and pathological processes
- VEGF −634 G>C may be a possible prognostic biomarker after bypass surgery.
- VEGF or KDR polymorphisms can be used as prognostic factors after revascularization surgery

PLOS One 10:e47158, 2012

VEGF and KDR

- Vascular endothelial growth factor (VEGF) is involved in vasculogenesis and vascular permeability in various intracranial lesions.
- In ischemic disease, cerebral angiogenesis is caused by the release of VEGF.
- VEGF affects vasculogenesis, endothelial cell proliferation and migration, vascular permeability, and stromal degradation through the activation of proteolytic enzymes that are involved in angiogenesis.
- VEGF binds its receptor tyrosine kinases, VEGF receptor-1 and VEGF receptor-2 (also known as kinase insert domain containing receptor, or KDR).
- KDR is the key receptor mediating angiogenesis and is essential for endothelial cell survival and integrity

PLOS One 10:e47158, 2012

VEGF in MMD

- Takekawa et al. reported increased VEGF expression in autopsy specimens from adults with moyamoya disease.
- Sakamoto et al. reported that the total meningeal cellularity and VEGF expression in the moyamoya dura was significantly higher in moyamoya patients compared to controls.
- Vascular endothelial growth factor A (VEGF-A) is a disulfide bonded dimeric glycoprotein that is a member of a protein family that includes VEGF-B, VEGF-C, VEGF-D, and placental growth factor (PGF). The gene that encodes VEGF-A is located on human chromosome 6 and comprises a 14-kb coding region with eight exons.
- VEGF acts on endothelial cells particularly through Flt-1 and KDR.
- Epidermal growth factor (EGF) arising from hypoxia stimulates Flt-1 expression and inhibits KDR expression.

PLOS One 10:e47158, 2012

Research paper

Plasma matrix metalloproteinases, cytokines and angiogenic factors in moyamoya disease

Table 1 Plasma cytokine assays in moyamoya disease patients and controls

Cytokines	Control Mean±SEM	Moyamoya disease Mean±SEM	p Value
MMP-1	149.42±25.89 pg/ml	392.84±118.20 pg/ml	0.2300†
MMP-2	52.29±4.32 ng/ml	59.83±3.21 ng/ml	0.2159†
MMP-3	13.59±1.78 ng/ml	1.76±0.38 ng/ml	<0.0001*
MMP-9	3.53±1.34 ng/ml	115.53±23.43 ng/ml	0.0002‡
MMP-12	OOR below	8.92±0.90 pg/ml	NA
TIMP-1	62.02±6.15 ng/ml	36.68±7.18 ng/ml	0.0180†
TIMP-2	107.96±5.74 ng/ml	93.27±3.46 ng/ml	0.0460†
MCP-1	79.90±24.86 pg/ml	1205.61±201.11 pg/ml	<0.0001‡
Resistin	3.27±0.65 ng/ml	3.38±0.44 ng/ml	0.8960†
IL-1β	0.19±0.06 pg/ml	0.70±0.13 pg/ml	0.0315†
IL-6	1.78±0.35 pg/ml	1.75±0.25 pg/ml	0.9513†
IL-8	OOR below	6.13±1.77 pg/ml	NA
TNFα	12.15±5.00 pg/ml	7.77±3.02 pg/ml	0.2030*
VEGF	24.22±7.18 pg/ml	482.09±120.77 pg/ml	0.0018‡
PDGF-BB	112.75±33.34 pg/ml	2071.48±673.75 pg/ml	0.0123‡

J Neurol Neurosurg Psychiatry 2010;81:673e678

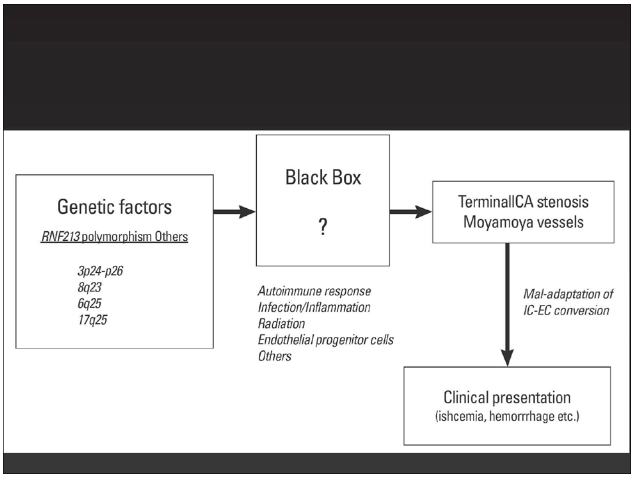
JoS JOURNAL OF STROKE

Journal of Stroke 2014;16(2):65-72
http://dx.doi.org/10.5853/jos.2014.16.2.65

Review

Genetics and Biomarkers of Moyamoya Disease: Significance of *RNF213* as a Susceptibility Gene

- The RNF213 gene (RNF213) in the 17q25-ter region was identified as a novel susceptibility gene for MMD among East Asian population
- A polymorphism in c.14576G>A in RNF213 was identified in 95% of familial patients with MMD and 79% of sporadic cases
- RNF213 was found to correlate with the early-onset and severe forms of MMD



진단

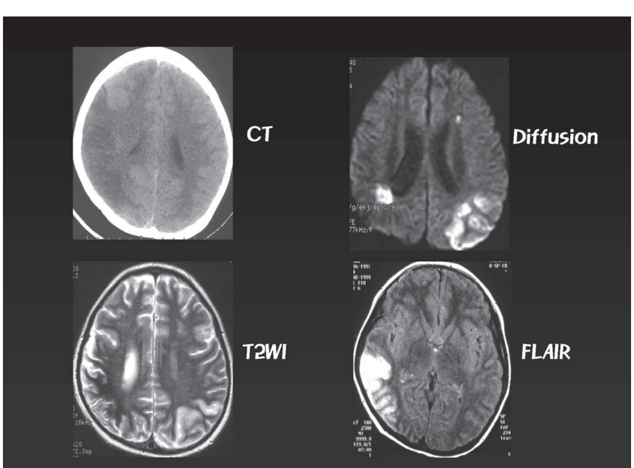
1. 병력
2. 뇌 CT
   Type I, II : 정상
   Type III, IV, V: 정상, 다발성 뇌경색, 뇌위축,
3. MRI : 특수 검사 및 전문의 판독
4. 혈관조영술, MR을 이용한 조영술
5. 뇌파검사
6. 혈류 검사 (SPECT)
7. MR 특수 검사: Diffusion, Perfusion, FLAIR image for infarction
9. PET, TCD

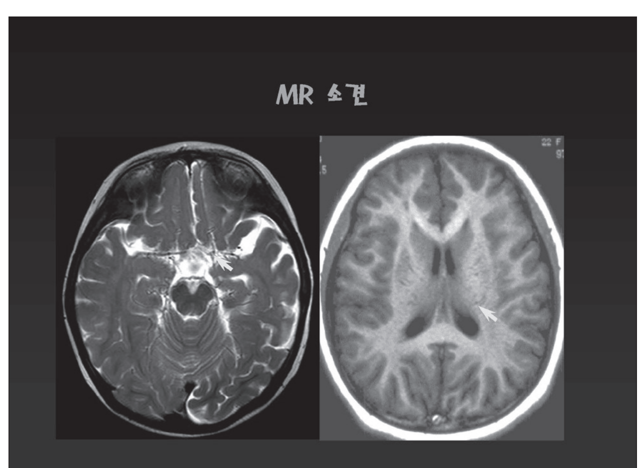
병력

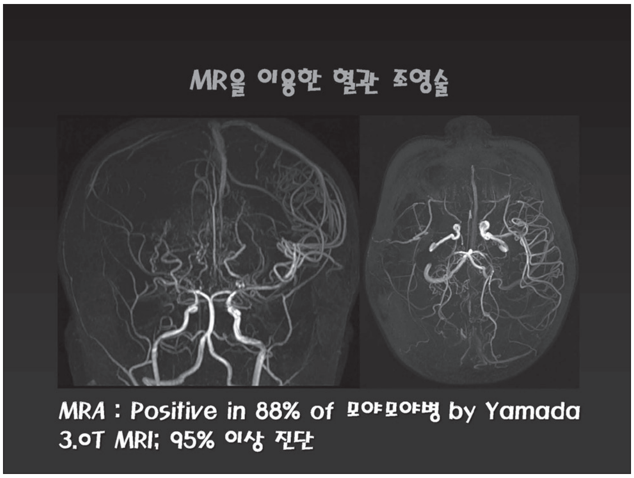
1. 일시적 허혈 증상 또는 뇌경색
   - 일시적 허혈을 유발하는 인자 : 울고 나서, 기침 후, 악기 불다가, 노래 부르다가, 뜨거운 음식 먹고 나서
2. 발작
3. 기타
   - 두통, 시력 저하, 감각 이상
   - 불수의 운동 (involuntary chorea movements)
   - 지능 저하

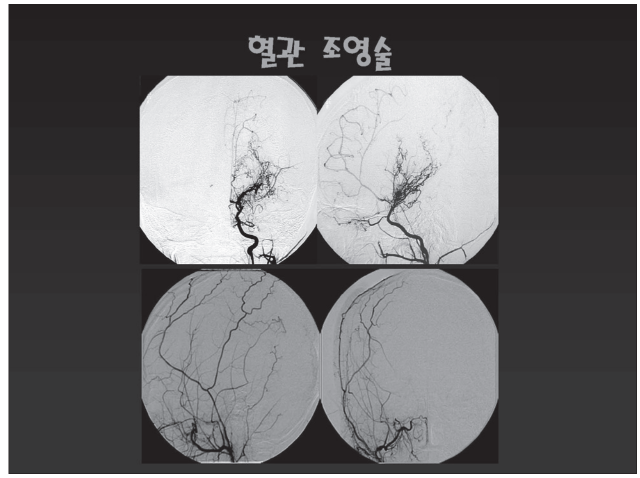
모야모야 환자에서 두통

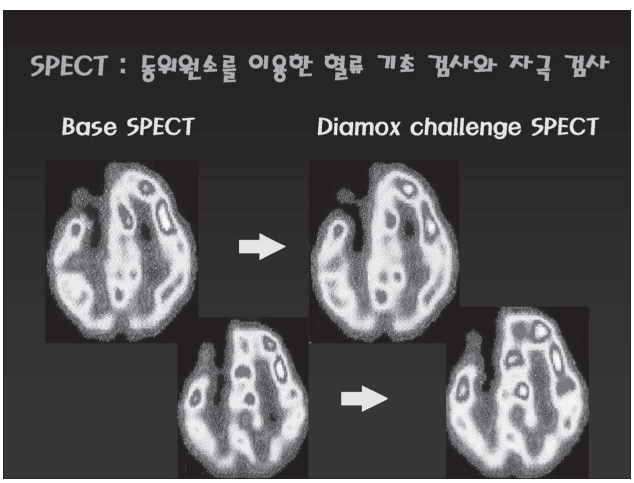
- ✓ 아침에 많다.
- ✓ 구토 동반
- ✓ 편두통과 유사한 안구통을 동반
- ✓ 약에 효과가 신통치 않다.
- ✓ 수 시간 동안 지속된다.
- ✓ 조절이 매우 어렵다
- ✓ 수술 후에도 상당 기간 지속이 되고, 더 심해지는 경우도 있다.

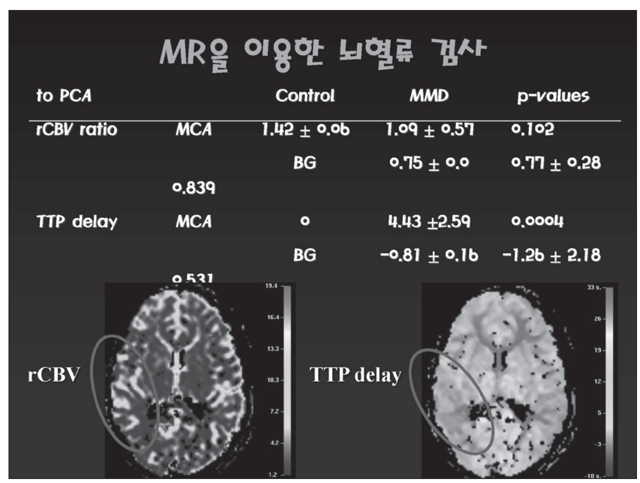


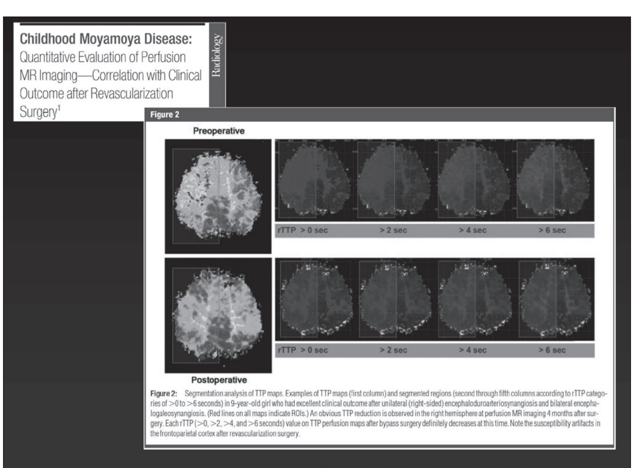


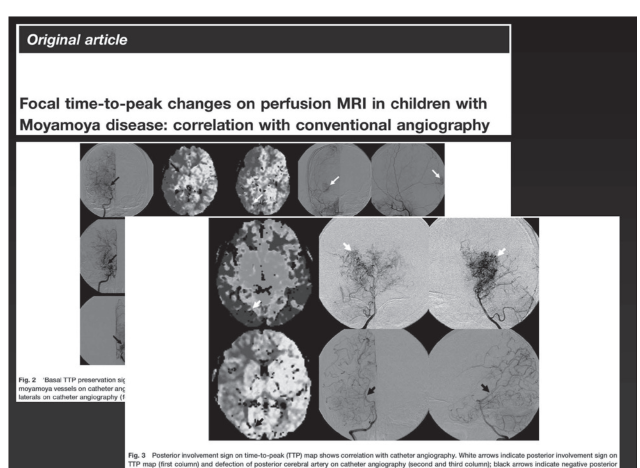

MR을 이용한 혈관 조영술
MRA : Positive in 88% of 모야모야병 by Yamada
3.0T MRI; 95% 이상 진단

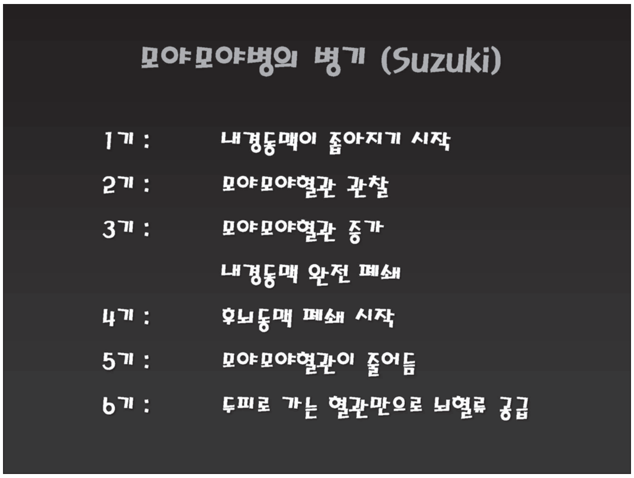

혈관 조영술

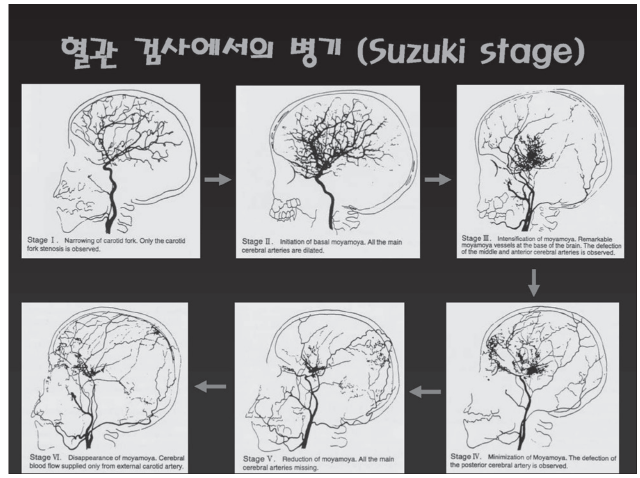

SPECT : 동위원소를 이용한 혈류 기초 검사와 자극 검사
Base SPECT
Diamox challenge SPECT

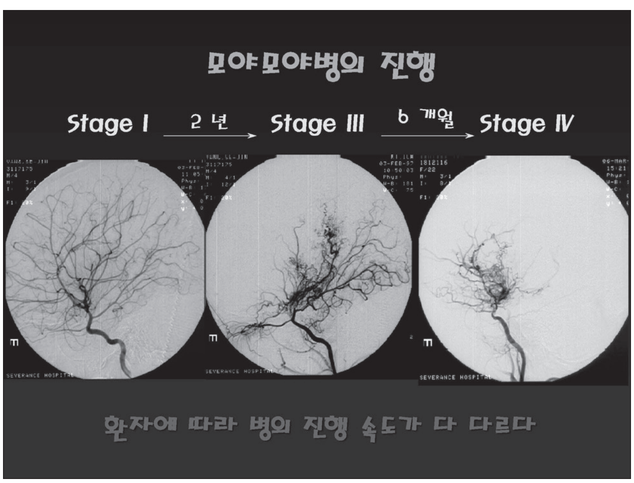

MR을 이용한 뇌혈류 검사
to PCA
Control
MMD
p-values
rCBV ratio
MCA
1.42 ± 0.06
1.09 ± 0.57
0.102
BG
0.75 ± 0.0
0.77 ± 0.28
0.839
TTP delay
MCA
0
4.43 ±2.59
0.0004
BG
-0.81 ± 0.16
-1.26 ± 2.18
0.531
rCBV
TTP delay

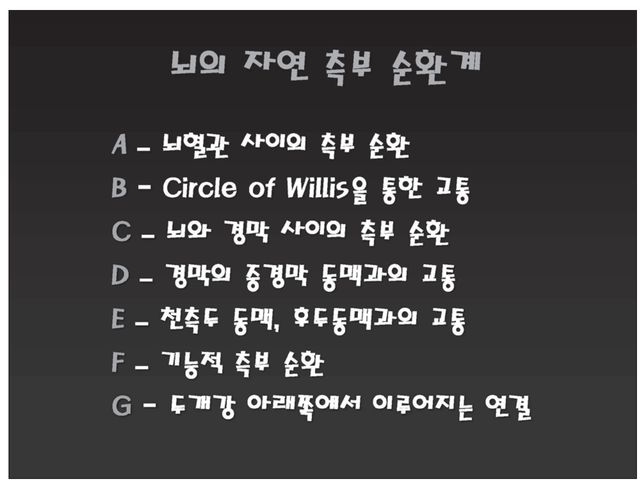

Childhood Moyamoya Disease:
Quantitative Evaluation of Perfusion MR Imaging—Correlation with Clinical Outcome after Revascularization Surgery[1]
Radiology
Figure 2
Preoperative
Postoperative
Radiology: Volume 251: Number 1—April 2009

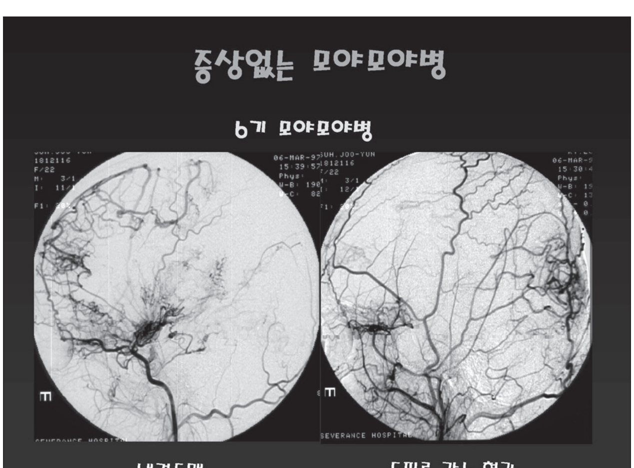

Original article
Focal time-to-peak changes on perfusion MRI in children with Moyamoya disease: correlation with conventional angiography


모야모야병의 병기 (Suzuki)
1기 : 내경동맥이 좁아지기 시작
2기 : 모야모야혈관 관찰
3기 : 모야모야혈관 증가
내경동맥 완전 폐쇄
4기 : 후뇌동맥 폐쇄 시작
5기 : 모야모야혈관이 줄어듬
6기 : 두피로 가는 혈관만으로 뇌혈류 공급

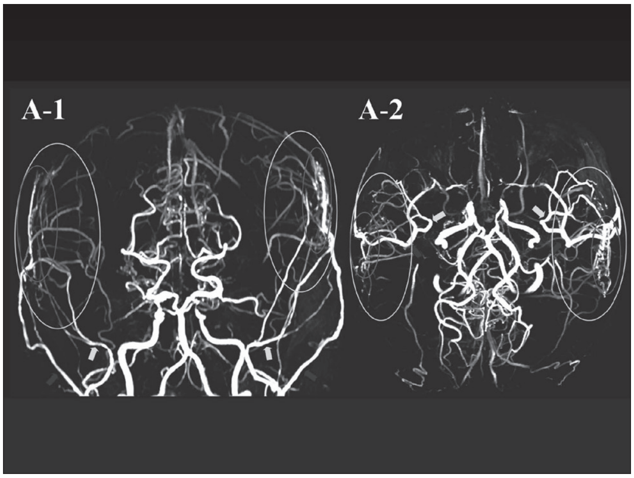

혈관 검사에서의 병기 (Suzuki stage)
Stage Ⅰ. Narrowing of carotid fork. Only the carotid fork stenosis is observed.
Stage Ⅱ. Initiation of basal moyamoya. All the main cerebral arteries are dilated.
Stage Ⅲ. Intensification of moyamoya. Remarkable moyamoya vessels at the base of the brain. The defection of the middle and anterior cerebral arteries is observed.
Stage Ⅳ. Minimization of Moyamoya. The defection of the posterior cerebral artery is observed.
Stage Ⅴ. Reduction of moyamoya. All the main cerebral arteries missing.
Stage Ⅵ. Disappearance of moyamoya. Cerebral blood flow supplied only from external carotid artery.

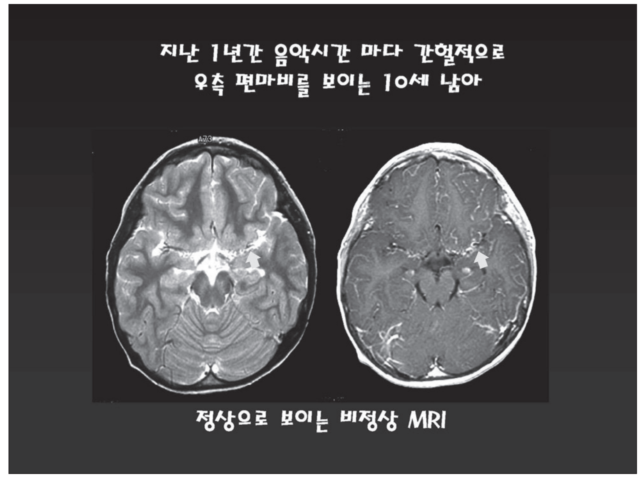

모야모야병의 진행
Stage I
2 년
Stage III
6 개월
Stage IV
환자에 따라 병의 진행 속도가 다 다르다

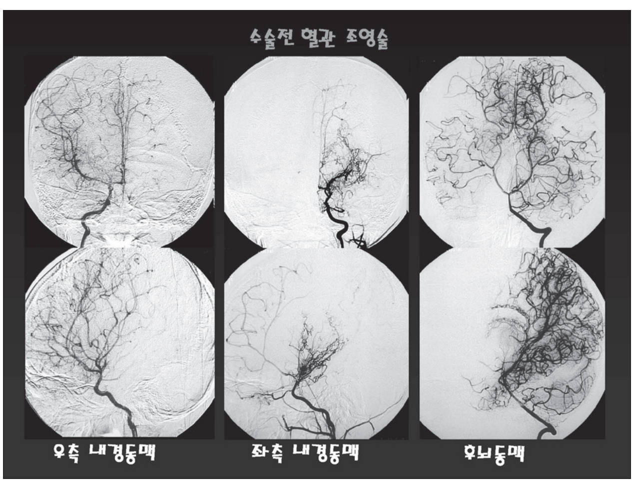

뇌의 자연 측부 순환계
A – 뇌혈관 사이의 측부 순환
B – Circle of Willis을 통한 교통
C – 뇌와 경막 사이의 측부 순환
D – 경막의 중경막 동맥과의 교통
E – 천측두 동맥, 후두동맥과의 교통
F – 기능적 측부 순환
G – 두개강 아래쪽에서 이루어지는 연결

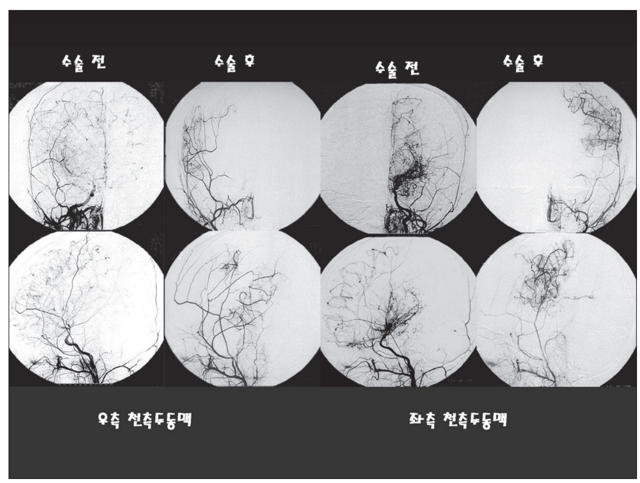

증상없는 모야모야병
6기 모야모야병
내경동맥
두피로 가는 혈관


모야모야병의 병기에 따른 수술 결정
1기 : 내경동맥이 좁아지기 시작
2기 : 모야모야혈관 관찰
3기 : 모야모야혈관 증가
내경동맥 완전 폐쇄
4기 : 후뇌동맥 폐쇄 시작 – 3 차 수술??
5기 : 모야모야혈관이 줄어듬 – 수술 X
6기 : 두피로 가는 혈관만으로 뇌혈류 공급 – 수술 XXX









ORIGINAL ARTICLE

Pediatric Moyamoya Disease:
An Analysis of 410 Consecutive Cases

TABLE 3: Surgical Outcomes of 410 Children with Moyamoya Disease

Parameters	Number of Cases (%)
Overall clinical outcome (n = 410 patients)	
Excellent	270 (66)
Good	63 (15)
Fair	62 (15)
Poor	15 (4)
MRI data (n = 393 patients)	
Favorable	295 (75)
Unfavorable	98 (25)
Extent of revascularization of MCA territory (n = 526 hemispheres)	
Good	177 (34)
Fair	294 (56)
Poor	55 (10)
Extent of revascularization of ACA territory (n = 494 hemispheres)	
Good	216 (44)
Fair	219 (44)
Poor	59 (12)
Extent of revascularization of PCA territory (n = 29 hemispheres)	
Good	20 (69)
Poor	9 (31)
Perfusion MRI data (n = 185 patients)	
Favorable	121 (65)
Unfavorable	64 (35)
SPECT changes (n = 355 patients)	
Favorable	214 (60)
Unfavorable	141 (40)
Neuropsychological test (n = 106 patients)	
IQ ≥90	71 (67)
IQ <90	35 (33)

MRI = magnetic resonance imaging; MCA = middle cerebral artery; ACA = anterior cerebral artery; PCA = posterior cerebral artery; SPECT = single-photon emission computerized tomography; IQ = intelligence quotient.

ANN NEUROL 2010;68:92–101

임상 증상에 따른 수술 결정

소아 : 뇌허혈성, 반복, 진행이 아주 빠름

성인 : 뇌출혈성, 1회성, 진행이 아주 느림

증상 유발 원인

1) 내경동맥의 폐쇄 속도
2) 측부 순환 형성 속도
3) 뇌의 혈류 필요량

편측 모야모야병

	편측 비율	평균 나이	환자 수	남 : 여	혈관 검사 소견	평균 추적 기간	반대측 진행	진행까지 기간 (개월)
Kelly et al., 2006	17.8% (28/157)	29.8	5	1 : 4	Normal (n=4) or only ACA abnormality (n=1)	25?	2/5 (40%)	13.5 (8-19)
Seol et al., 2006	3.4% (7/204)	5.1	7	2 : 5	Normal (n=4) or only ACA abnormality (n=3)	65	2/7 (29%)	29.5 (27-32)
Smith et al., 2008	14% (33/235)	10.4	29	14 : 15	Normal (n=12) or some abnormalities (n=21) ?	67	8/29 (28%)	26.4 (12-102) ?
Kim et al., 2010	12.9% (53/410)	?	?		Normal (n=33) or ACA abnormallity (n=20)	?	24/53 (45%)	23 (0.5-150)
Park et al., 2011	15.4% (40/259)	8.7	34	18:16	Normal (n=12) or ACA or MCA abnormal (n=22)	35.3	20/34 (58.8%)	17.3(7.3-34.8)
Yeon et al., 2011	11.4% (45/394)	9.9	45	24 : 21	Normal (n=15) or only ACA abnormality (n=30)	53	8/45 (18%)	31.3 (21-71)

Childs Nerv Syst (2011) 27:1281–1287
DOI 10.1007/s00381-011-1469-y

ORIGINAL PAPER

Natural history and progression factors of unilateral moyamoya disease in pediatric patients

Eun Kyung Park · Yun-Ho Lee · Kyu-Won Shim · Joong-Uhn Choi · Dong-Seok Kim

Table 1 Summary of patient demographics and clinical characteristics

Total numbers	34
M/F ratio	18:16
Age (yrs)	4.1–19.9 (mean, 8.7 years)
Follow-up periods (months)	18.1–100 (mean, 35.3 months)
Family history	6 (17.6%)
Presentation symptoms	
Transient ischemic attack	23 (67.6%)
Infarction	4 (11.8%)
Headache	6 (17.6%)
Incidental	1 (3.0%)

Table 2 Clinical characteristics of the progression group

Case	Sex	Age at diagnosis	Baseline abnormality		Family history	Initial Suzuki grade and side	Bilateral EDAS
			ACA	MCA			
2	F	17				4/R	+
3	M	9			+	3/R	+
4	F	14	+			3/R	
6	M	6				3/R	
7	M	11	+			3/R	
8	F	7	+	+		3/L	
10	F	9	+		+	4/R	
11	M	8			+	3/R	
12	M	4	+			3/L	
13	F	7	+			3/R	+
14	F	5	+	+		3/R	+
19	F	8	+			3/R	+
21	F	8				3/L	+
22	F	8		+	+	2/R	
25	F	8	+	+	+	4/L	+
28	F	14	+		+	2/R	
29	M	6	+			2/L	
32	M	13	+			2/L	
33	M	10	+			3/L	
34	F	14	+	+		2/L	

	Non-progression group	Progression group
Numbers	14 (41.2%)	20 (58.8%)
Age (mean)	4–20 (mean, 8.8 years)	4–17 (mean, 9.3 years)
Sex (M/F)	10:4 (2.5:1)	8:12 (1:1.5)
Initial ACA/MCA abnormality	7 (50%)	15 (75%)
Family history	0/6 (0%)	6/6 (100%)

ELSEVIER

Clinical Neurology and Neurosurgery 99 Suppl. 2 (1997) S11–S18

Clinical Neurology and Neurosurgery

Natural history of Moyamoya disease: comparison of activity of daily living in surgery and non surgery groups

Joong Uhn Choi *, Dong Seok Kim, Eun Young Kim, Kyu Chang Lee

Department of Neurosurgery, Brian Research Institute, Yonsei University College of Medicine, Seoul, South Korea

추적기간 12 ~ 216 개월 (평균 67.2 개월)

수술 하지 않은 환자

ADL changes of non-surgery group at different age and

Change of ADL	Age (years)	
	<6	6–15
Improvement	3	2 (1[a])
No change	3	1
Aggravation	9	3
No. of patients	15	6 (1[a])

수술 받은 환자

ADL changes of the patients surgically treated at different age

Change of ADL	Age group (years)	
	<6	6–15
Improvement	13	4
No change	3	3
Aggravation	1	2
Total	17	9

Pediatric Neurology

Volume 1, Issue 5, September–October 1985, Pages 274–277

ELSEVIER

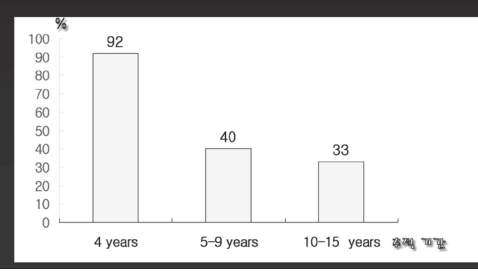
Original article

Prognosis of occlusive disease of the circle of Willis (moyamoya disease) in children *

Toru Kurokawa, MD *, Shigeru Tomita, MD*, Kohji Ueda, MD*, Osamu Narazaki, MD§, Toshio Hanai, MD§, Kanehiro Hasuo, MD†, Toshio Matsushima, MD‡, Katsutoshi Kitamura, MD‡

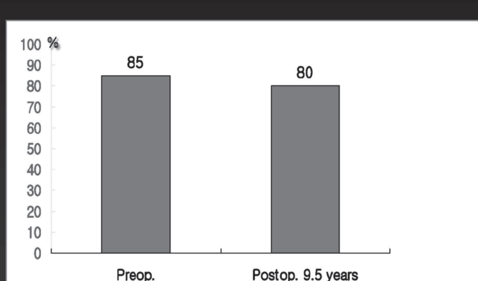
27 소아 환자, 수술 않고 보존적 치료, 1971 - 1984

정상 IQ (> 86)



정상 IQ 비율

Matsushima Y et al, 1997



Comparison of Neuropsychological variables to Clinical and Non-clinical control group in Moyamoya Disease

76 children with MMD; boys 31:girls 45, mean age=8.04 years
46 Attention-Deficit Hyperactivity Disorder; boys 36:girls 10, mean age=8.43 years
34 Normal control group; boys 19:girls 10, mean age=7.85 years

		MMD (N=76)	ADHD Clinical Control (N=46)	Normal Control (N=34)	Sig.	Post Hoc
Intelligence	Full Scale IQ	86.67	104.73	110.03	.000**	A < B,C
	Verbal IQ	92.07	107.02	112.70	.000**	A < B,C
	Performance IQ	80.71	100.93	103.91	.000**	A < B,C
	Verbal Comprehension	95.73	109.32	108.00	.001**	A < B,C
	Perceptual Organization	87.03	102.42	106.92	.000**	A < B,C
	Concentration	84.30	97.27	111.92	.000**	A < B,C
	Performance Speed	93.93	95.93	104.70	.019*	A < C
Memory	MQ	87.48	97.72	101.75	.014*	A < B,C

Severance Children's Hospital

Comparison of Neuropsychological variables to Clinical and Non-clinical control group in Moyamoya Disease

76 children with MMD; boys 31:girls 45, mean age=8.04 years
46 Attention-Deficit Hyperactivity Disorder; boys 36:girls 10, mean age=8.43 years
34 Normal control group; boys 19:girls 10, mean age=7.85 years

		MMD (N=76)	ADHD Clinical Control (N=46)	Normal Control (N=34)	Sig.	Post Hoc
Attention	Inattention	79.95	68.91	78.84	.363	
	Impulsivity	85.84	73.95	66.58	.118	
	Mean of Response Time	55.24	52.52	50.11	.452	
	Standard Deviation of Response Time	87.32	78.12	73.16	.264	
Executive Function	Perseverative errors	32.71	42.21	51.73	.164	
	Conceptualization	22.71	31.18	45.10	.070	
	Category completed	26.20	35.31	50.00	.006**	A < C
	Learn to Learn	18.86	34.76	42.10	.006**	A < B,C

Severance Children's Hospital

Qualitative rate of Abnormal range in Moyamoya Disease.

		N (tot. N)	Mean(SD)	% in below average or abnormal range
Intelligence	Full Scale IQ	14 (76)	86.67	30.4
	Verbal IQ	9 (76)	92.07	19.6
	Performance IQ	15 (76)	80.72	32.6
Memory	MQ	10 (51)	87.48	32.3
Attention	Inattention	23 (68)	79.95	49.0*
	Impulsivity	21 (68)	85.84	44.7*
	Mean of Response Time	13 (63)	55.24	39.4
	SD of Response Time	26 (68)	87.32	55.3*
Executive Function	Perseverative errors	6 (51)	32.71	28.6
	Conceptualization	12 (51)	22.71	57.1*
	Category completed	6 (51)	26.19	28.6
	Learn to Learn	11 (51)	18.86	52.4*

Severance Children's Hospital

Cerebrovascular Diseases

Original Paper

Cerebrovasc Dis 2011;31:230–237
DOI: 10.1159/000321901

Received: July 30, 2010
Accepted: September 17, 2010
Published online: December 21, 2010

Neurocognitive Profiles of Children with Moyamoya Disease before and after Surgical Intervention

Ji Yeoun Lee[a] Ji Hoon Phi[a] Kyu-Chang Wang[a] Byung-Kyu Cho[a]
Min-Sup Shin[b] Seung-Ki Kim[a]

Table 3. Preoperative and postoperative scores on various neurocognitive tests (means ± SD)

	Preoperative (n = 65)	Postoperative (n = 65)	Paired t test
Intellectual functioning			
FSIQ	107.2 ± 14.2	108.1 ± 12.7	–0.79 (0.44)
VIQ	107.8 ± 13.2	105.8 ± 12.7	1.59 (0.18)
PIQ	105.3 ± 16.1	109.0 ± 13.11	–2.76 (0.01)

- 수술 전 뇌경색이 없어야 좋은 결과를 기대할 수 있다.
- 수술로 인한 혈류 개선이 인지 기능 개선이나 유지에 도움이 된다.
- 병이 진행하기 전에 적극적 치료를 하는 것이 중요하다.

Child's Nervous System
https://doi.org/10.1007/s00381-018-3804-z

ORIGINAL PAPER

Neuropsychological impacts of indirect revascularization for pediatric moyamoya disease

Table 3 Preoperative and postoperative scores on neuropsychological tests (means + SD)

	Pre (SD) ($n = 55$)	Post (SD) ($n = 55$)	df	Paired t test	p value
K-WISC-III					
FSIQ	98.9 (15.3)	103.3 (17.1)	54	− 3.073	.003*
VIQ	103.5 (13.1)	103.8 (15.2)	54	− .285	.777
PIQ	93.5 (17.6)	101.9 (19.8)	54	− 4.475	.000*
Verbal comprehension (VC)	104.0 (13.0)	104.0 (14.9)	52	− .028	.978
Perceptual organization (PO)	93.6 (16.3)	103.7 (18.2)	52	− 5.169	.000*
Freedom from distractibility (FD)	95.6 (15.6)	98.2 (14.1)	52	− 1.536	.130
Processing speed (PS)	99.4 (19.6)	99.0 (19.5)	52	.203	.840
Rey-Kim memory test					
MQ	95.3 (17.7)	105.4 (14.3)	45	− 4.021	.000
Verbal delayed recall	9.3 (3.4)	9.4 (2.8)	38	− .190	.850
Verbal delayed recognition	8.9 (2.7)	10.3 (1.9)	37	− 2.972	.005
Visuospatial/constructional	9.9 (3.9)	11.2 (3.6)	38	− 2.124	.040
Visual immediate recall	9.4 (3.9)	13.2 (7.0)	38	− 3.347	.002
Visual delayed recall	9.6 (4.2)	13.2 (10.5)	37	− 2.148	.038
Children's Color Trails Test (CCTT)					
CCTT-1 (s)	25.4 (14.9)	20.0 (9.6)	49	3.836	.000
CCTT-2 (s)	57.3 (26.2)	45.1 (20.6)	49	3.689	.001
CCTT-1 T score	46.6 (12.7)	50.6 (11.0)	49	− 2.307	.025
CCTT-2 T score	46.3 (13.1)	51.2 (12.4)	49	− 2.113	.040
Wisconsin Card Sorting Test (WCST)					
Perseverative errors	49.8 (11.5)	56.3 (9.1)	22	− 3.294	.003
Non-perseverative errors	45.4 (12.8)	57.0 (8.0)	22	− 4.729	.000
Conceptual level responses	47.0 (13.1)	58.3 (10.6)	22	− 5.563	.000
Categories completed	4.3 (2.2)	5.7 (.8)	22	− 3.364	.003
Learning to learn	− 1.8 (2.4)	− 1.6 (1.9)	3	− .161	.883
Trials to complete 1st category	13.9 (6.8)	15.1 (9.4)	9	− 1.276	.234
Failure to maintain set	.8 (1.1)	.9 (.8)	20	− .336	.741
Advanced Test of Attention (ATA)—visual only administered					
Inattention	68.6 (19.6)	61.2 (18.4)	46	2.437	.019
Impulsivity	71.9 (24.2)	61.1 (21.5)	49	4.029	.000
Reaction time (RT)	53.9 (10.3)	53.2 (13.2)	49	.328	.745
RT SD	65.4 (21.1)	57.2 (18.2)	49	2.253	.029

소아 모야모야병 요약

- 병의 경과를 바꿀 수 있는 약물 치료는 없다.
- 치료하지 않은 소아 환자는 혈관 상태나 임상 상태가 결국은 악화한다.
- 5년 내에 60-70%의 소아 환자는 증상 진행을 보인다.
- 치료를 하지 않으면 소아에서 결과는 대체로 나쁘다.
- 치료 당시의 신경학적 상태가 치료의 장기 결과를 반영한다.
- 조기 진단, 조기 치료가 중요하다.

감사합니다.

Diagnostic Image of Moyamoya Disease: Radiologic Image & Differential Diagnosis of Moyamoya Disease

김용배

연세대

Moyamoya disease (MMD) can be defined as a progressive steno-occlusive disease at terminal portions of the bilateral internal carotid arteries and their proximal branches with prominent collateral artery formation. Prompt diagnosis and accurate assessment could significantly improve the prognosis of MMD. To date, different imaging modalities has been used for diagnosis of MMD, including digital subtraction angiography (DSA), computed tomography (CT), magnetic resonance imaging (MRI), single-photon emission CT (SPECT), and positron emission tomography (PET). All of these imaging modalities are useful in certain situation where the specific information of MMD is in need upon their quantitative or qualitative evaluation. Furthermore, recent reveal of superiority of bypass surgery in hemorrhagic MMD may provide us an upgraded understanding about the impact of angiographic feature and hemorrhagic site on the prognosis of hemorrhagic MMD.

Differential diagnoses of MMD may be including cerebral atherosclerosis, autoimmune diseases, meningitis, cerebrovascular damage after head injury, and many anomalies of the intracranial arteries. Additionally, radiation therapy, neurofibromatosis type 1, Down's syndrome, sickle cell disease, and even autoimmune diseases may develop a striate collateral network of the proximal MCA with a web-like pattern. Among them, an anomaly of the middle cerebral artery (MCA) is well mimicking MMD. Among the anomalies of the MCA, rete MCA (R-MCA) is a web-like anomaly of the MCA that does not coalesce and forms a prominent, large single branch from the plexiform vessels in the fetal stage.

In this review, the usefulness of each diagnostic modality are conveyed and discussed to estimate the current status and the prognosis of MMD so that the most suitable strategies could be yield for each individual patient. And, by reviewing different, moyamoya-mimicking disease, we hope valuable insights could be provided for vascular neurosurgeons to avoid misdiagnosis and unnecessary treatments.

Session II. Moyamoya Disease: Treatment

좌장: 성재훈 (가톨릭대), 고현송 (충남대)

1. Bypass strategies for Moyamoya disease: Risk and incidence of complication & avoidance of complication from revascularization surgery

   연제영 (성균관대)

2. Surgical role in hemorrhagic Moyamoya disease

   김정은 (서울대)

3. Endovascular treatment for targeted embolization in Moyamoya disease with hemorrhagic manifestation

   반승필 (서울대)

Bypass strategies for Moyamoya disease: Risk and incidence of complication & avoidance of complication from revascularization surgery

연제영

성균관대

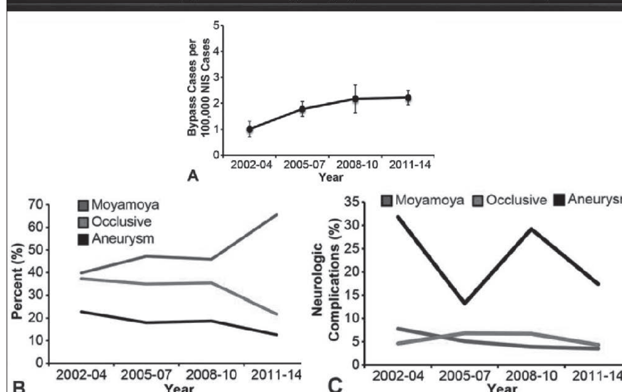
2019.9.7. SKEN-KSCVS 합동연수강좌 11:00-11:25

Bypass Strategies for Moyamoya Disease: Risk and Incidence of Complication and its Avoidance

Je Young Yeon

Department of Neurosurgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

Trends in Bypass Surgery (USA, 2002-2014)



Winkler et al., Neurosurg Focus, 2019

Bypass indications and Level of Evidence

- Flow-augmentation for **ischemic moyamoya disease**
  - level of evidence 4; grade of recommendation C
- Flow-augmentation for **hemorrhagic moyamoya disease**
  - level of evidence 1B; grade of recommendation A
- Flow-augmentation for **medially refractory carotid occlusion**
  - level of evidence 5; grade of recommendation D

Esposito et al., Acta Neurochir, 2018

Hemorrhagic MMD – JAM trial

- Multicentered, prospective, randomized, controlled trial conducted by 22 institutes in Japan
- Intracranial hemorrhage within the preceding year
- Conservative care or bilateral EC-IC direct bypass
- Rebleeding rate for 5 years (*P*=0.042 in the K-M analysis)
  - 5 patients in the surgical group (11.9%), 2.7%/year
  - 12 in the nonsurgical group (31.6%), 7.6 %/year

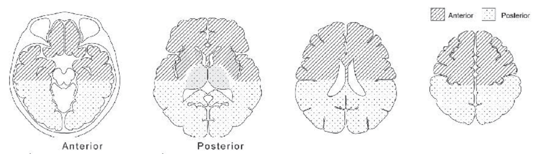
	Surgical Group (n=42)		Nonsurgical Group (n=38)			
	n	Rate, %	n	Rate, %	Hazard Ratio (95% CI)	*P* Value
Primary end point	6	14.3	13	34.2	0.391 (0.148–1.029)	0.057
Recurrent bleeding	5	11.9	12	31.6	0.355 (0.125–1.009)	0.052
Completed stroke	1	2.4	0	0.0	…	…
Crescendo TIA (bypass required)	0	0.0	1	2.6	…	…

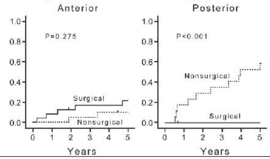
* Ischemic stroke or TIA : 0.5%/year in each group

Miyamoto et al., Stroke, 2014

Hemorrhagic MMD – JAM trial

- Analysis in the Japan Adult Moyamoya (JAM) Trial
- Hemorrhagic site classified as either anterior or posterior
- Diffuse IVH or SAH → anterior type





Patients with **posterior hemorrhage** are at **higher risk of rebleeding** and accrue **greater benefit from surgery**.

Takahashi et al., Stroke, 2016

Risk factors for postoperative stroke

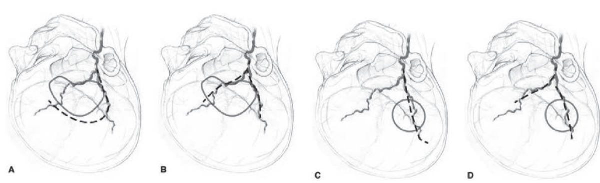
- Meta-analysis of 8 studies encompassing 1649 patients

- Preoperative ischemic event
- PCA involvement
- Diabetes

Preoperative possible risk factors	Number of articles	I^2	P value for heterogeneity	Pooled OR(95%CI)	P for pooled results	P for Begg's test
Ischemic events	5	74.4%	0.001*	1.40 (1.02–1.92)	0.039*	0.230
PCA involvement	5	83.4%	0.000*	2.64 (1.17–5.95)	0.019*	0.806
Suzuki stage	4	32.8%	0.215	1.20 (0.97, 1.49)	0.101	0.734
Surgery type (DB as reference)	4	0.0%	0.601	1.12 (1.03, 1.33)	0.017*	0.806
Age	5	38.2%	0.167	1.02 (1.00, 1.04)	0.090	0.806
Mae sex	4	0.0%	0.595	1.16 (0.75–1.82)	0.504	1.000
Diabetes	3	5.9%	0.345	4.02 (1.59, 10.16)	0.003*	0.296
Hypertension	2	0.0%	0.494	0.70 (0.31, 1.58)	0.392	–

Wei et al., BMC Neurology, 2019

Skin incision and wound complication



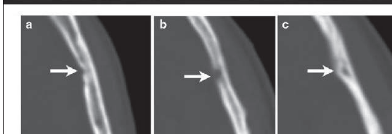
Procedure	No.	No. w/ Major WHD	%
Direct bypass via targeted craniotomy			
Linear	62	1	1.6
Incomplete Y	107	3	2.8
Direct bypass after previous hemicraniotomy	1	1	
Combined w/ EMS via pterional craniotomy			
Complete Y	42	6	14.3
Curved	24	1	4.2
Total	236	12	5.1

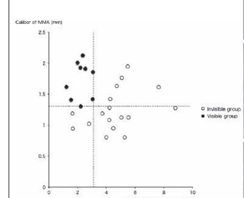
WHD = wound healing disorder.

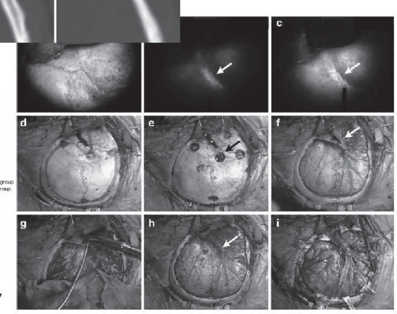
Major wound complication requiring revision

Acker et al., Neurosurg Focus, 2019

ICG angiography before craniotomy

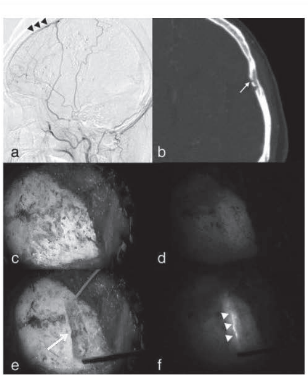






Tanabe et al., Acta Neurochir, 2017

ICG angiography during craniotomy



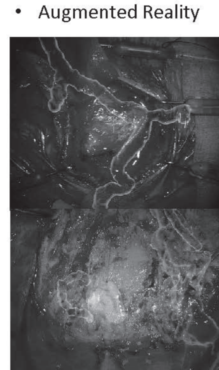
drilled the compact bone out with a cutting burr around the possible MMA course, which was estimated from the distance from the parietal branch of the superior temporal artery (Fig. 1e).

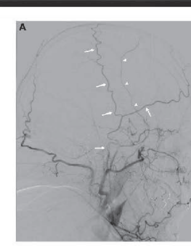
Then, the ICG was injected again, which showed the MMA course clearly (Fig. 1f). We could carry out a craniotomy posteriorly to the MMA and made an STA-MCA direct bypass.

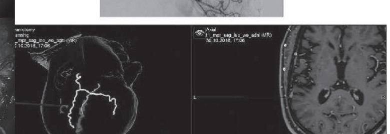
Kimura et al., Acta Neurochir, 2017

Preoperative planning

- Augmented Reality







Rychen et al., Operative Neurosurg, 2019

Surgical role in hemorrhagic Moyamoya disease

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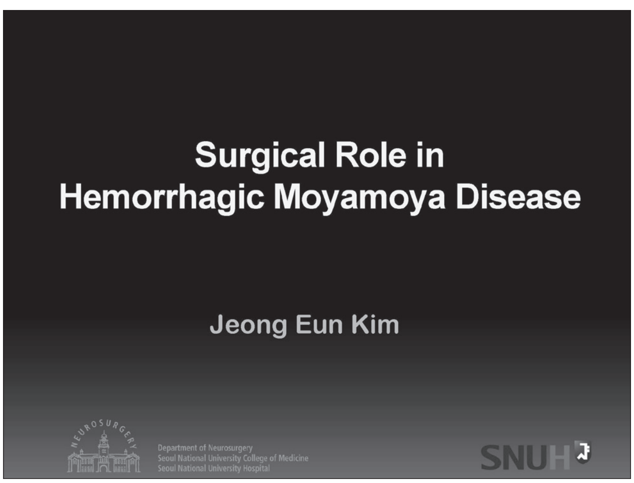
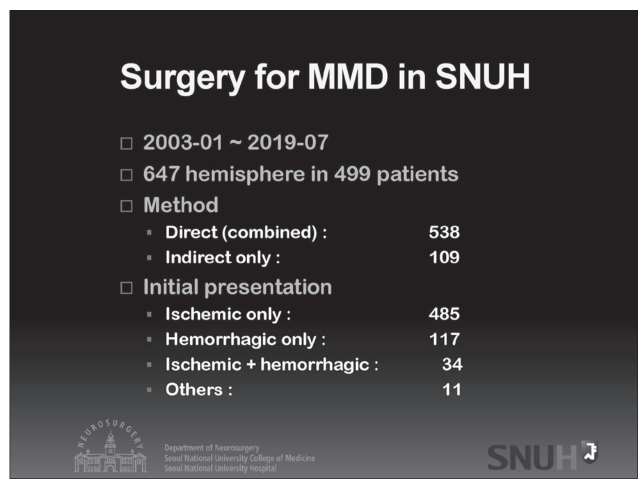
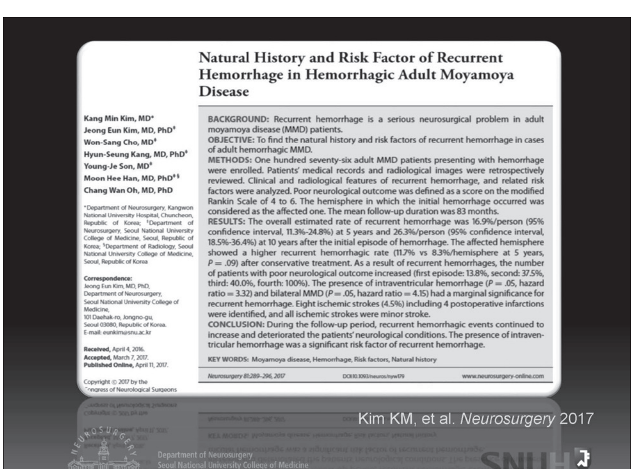
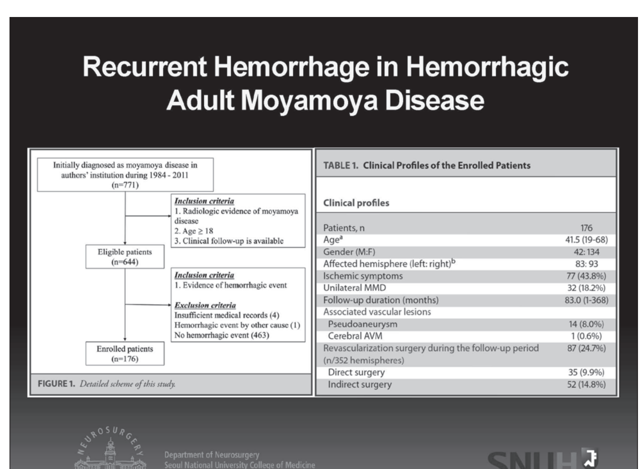
서울대









Recurrent Hemorrhage in Hemorrhagic Adult Moyamoya Disease

- Mean follow-up duration
  - 83 months (1-368)
- Recurrent hemorrhage
  - 45 times
- Recurrent hemorrhage rate
  - 16.9%/person at 5 year (3.4%/person-year)
  - 26.3%/person at 10 years

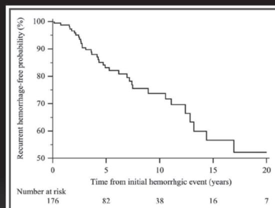


FIGURE 2. Overall recurrent hemorrhages in the enrolled patients.

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Recurrent Hemorrhage in Hemorrhagic Adult Moyamoya Disease

- Neurologic outcomes of patients

TABLE 2. Neurological Outcomes of Patients with Hemorrhagic Events at the Time of the Last Clinical Follow-up (n = 176)

No. of hemorrhagic event	No. of patients	No. of mRS ≥ 4 cases (%)	Mortality (%)
1	138	19(13.8)	1(0.7)
2	32	12(37.5)	4(12.5)
3	5	2(40.0)	1(20.0)
4	1	1(100.0)	1(100.0)

No., number; mRS, modified Rankin Scale.

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Recurrent hemorrhage in Hemorrhagic adult Moyamoya disease

- Risk factors of recurrent hemorrhages
  - IVH
  - Bilateral MMD

TABLE 4. Risk Factors of Recurrent Hemorrhage in Moyamoya Disease (Hemispheric Analysis)

	Univariate analysis[a] (P)	Multivariate analysis[b] (P)	Hazard ratio(95% CI)
Affected hemisphere	.06	.09	1.77(0.91-3.44)
Intraventricular hemorrhage	.04	.05	3.32(1.01-10.90)
Pseudoaneurysm	.04	.22	2.14(0.63-7.23)
Bilateral MMD	.04	.05	4.15(1.00-17.27)

MMD, moyamoya disease; CI, confidence interval.
[a]Kaplan–Meier analysis, log-rank test.
[b]Cox regression analysis.

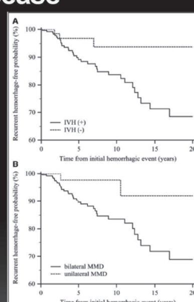

FIGURE 4. Risk factors of recurrent hemorrhage. A, Presence of IVH (P = .04, hazard ratio = 3.23 [95% CI, 1.50-6.97]). B, Bilateral MMD (P = .04, hazard ratio = 3.91 [95% CI, 1.71-8.94]).

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Revascularization Surgery During Follow-up

- 87 / 320 hemispheres (27%)

	combined	Indirect only
Ischemic-hemorrhagic	5	12
Hemorrhagic at first	30	40

- Recurrent hemorrhage after surgery
  - 1 in 30 combined
  - 5 in 40 indirect only

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Accompanying Lesions

- Pseudoaneurysm (n=14)
  - glue embolization : 5
  - conservative management : 9
    - Spontaneous regression : 4
    - Recurrent hemorrhage : 1
- 1 AVM : Gamma knife radiosurgery

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Ischemic Strokes After Initial Hemorrhag

- 8 ischemic strokes
  - 4 in observation group
  - 4 in surgery group (within 30 days after surgery) : 5.7%
  - All 8 ischemic strokes : minor stroke

  → Not disabling

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JAM (Japanese Adult Moyamoya) Trial

JAM Trial

- Multicenter randomized controlled trial
  - Hypothesis : **EC-IC bypass was beneficial** for **hemorrhagic** moyamoya disease
- Inclusion criteria
  - Intracranial hemorrhage within 12 months
  - Age : 16 – 65
  - mRS : 0-2, independent in daily life
  - Bilateral carotid lesions (unilateral excluded)
  - Staged bilateral surgery : STA-MCA anastomoses

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JAM Trial

- Miyamoto S, et al. *Stroke* 2014
- Takahashi JC, et al. *Stroke* 2016
- Funaki T, et al. *J Neurosurg* 2018 (1)
- Funkaki T, et al. *J Neurosurg* 2018 (2)
- Fujimura M, et al. *J Neurosurg* 2018
- Funaki T, et al. *J Neurosurg* 2019

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Effects of Extracranial–Intracranial Bypass for Patients With Hemorrhagic Moyamoya Disease
Results of the Japan Adult Moyamoya Trial

Susumu Miyamoto, MD, PhD; Takashi Yoshimoto, MD, PhD; Nobuo Hashimoto, MD, PhD; Yasushi Okada, MD, PhD; Ichiro Tsuji, MD, PhD; Teiji Tominaga, MD, PhD; Jyoji Nakagawara, MD; Jun C. Takahashi, MD, PhD; on behalf of the JAM Trial Investigators

Miyamoto S, et al. *Stroke* 2014

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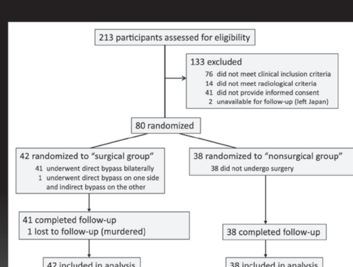
Effects of Extracranial-Intracranial Bypass for Patients with Hemorrhagic Moyamoya disease

- 2001-01 ~ 2008-06
- 80 patients
  - Surgical 42
  - Nonsurgical 38



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Effects of Extracranial-Intracranial Bypass for Patients with Hemorrhagic Moyamoya disease

- Primary end points : all adverse event
  - Rebleeding attack
  - Completed stroke resulting in significant morbidity
  - Significant morbidity or mortality from any medical cause
  - Requirement for bypass surgery for a nonsurgical patients
- Secondary end point
  - Rebleeding attack

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Effects of Extracranial-Intracranial Bypass for Patients with Hemorrhagic Moyamoya disease

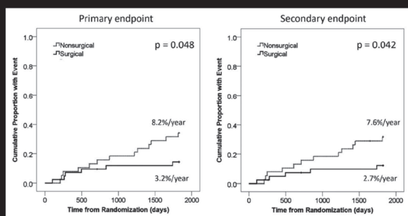

Figure 2. Kaplan-Meier cumulative curves for end points.

- Perioperative complications
  - 42 patients for 84 surgical procedures
  - 8 cases (9.5%)
    - Symptoms of local hyperperfusion : 3
    - TIA, Seizure, Local vasogenic edema, Scalp sore, Tear of a subcutaneous drainage tube

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Effects of Extracranial-Intracranial Bypass for Patients with Hemorrhagic Moyamoya disease

- Conclusion
  - Statistically marginal
  - Direct bypass surgery for adult patients with hemorrhagic moyamoya disease
    - → Reduces the rebleeding rate
    - → Improves a patient's prognosis

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Significance of the Hemorrhagic Site for Recurrent Bleeding
Prespecified Analysis in the Japan Adult Moyamoya Trial

Jun C. Takahashi, MD, PhD; Takeshi Funaki, MD, PhD; Kiyohiro Houkin, MD, PhD; Tooru Inoue, MD, PhD; Kuniaki Ogasawara, MD, PhD; Jyoji Nakagawara, MD, PhD; Satoshi Kuroda, MD, PhD; Keisuke Yamada, MD, PhD; Susumu Miyamoto, MD, PhD; on behalf of the JAM Trial Investigators*

Takahashi J, et al. *Stroke* 2016

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Significance of the Hemorrhagic Site for Recurrent Bleeding

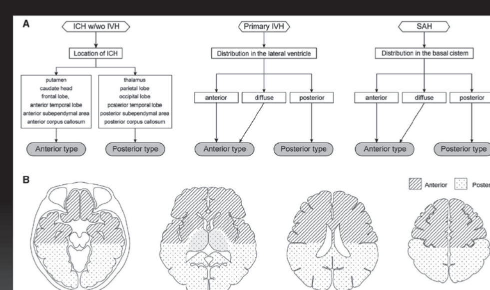

Figure 1. A, Flow diagram showing the algorithm for classifying hemorrhagic sites. B, Schematic illustrations showing topographical definitions of hemorrhagic sites. ICH indicates intracerebral hemorrhage; IVH, intraventricular hemorrhage; and SAH, subarachnoid hemorrhage.

Significance of the Hemorrhagic Site for Recurrent Bleeding

- Primary end points
  - All adverse effect including rebleeding attack
- Secondary end point
  - Rebleeding attack

Table 1. Primary End Point in Subgroups

Stratification Variable	Allocation: Nonsurgical (n=38)	Surgical (n=42)	*P* Interaction
Hemorrhagic site			0.013
Anterior			
No. of participants	21	24	
Primary end point (no. [%])	3 (14.3)	5 (20.8)	
HR (95% CI)	1.00 (reference)	1.62 (0.39–6.79)	
Posterior			
No. of participants	17	18	
Primary end point (no. [%])	10 (58.8)	1 (5.6)	
HR (95% CI)	1.00 (reference)	0.07 (0.01–0.55)	

Significance of the Hemorrhagic Site for Recurrent Bleeding

- HR for surgical group relative to the nonsurgical group
  - 0.07 for the posterior group

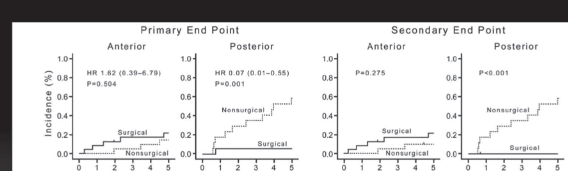

Figure 2. Kaplan–Meier estimates of the incidence of primary and secondary end points by hemorrhagic site. *P*=0.013 for interaction in the primary end point. Calculation of the hazard ratio (HR) and interaction *P* value was impossible for the secondary end point.

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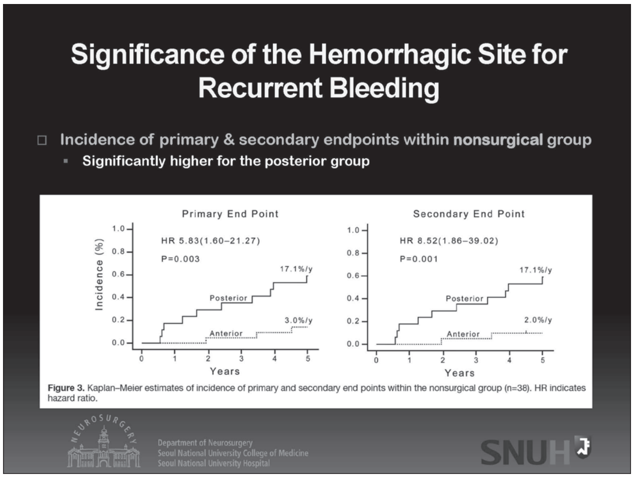

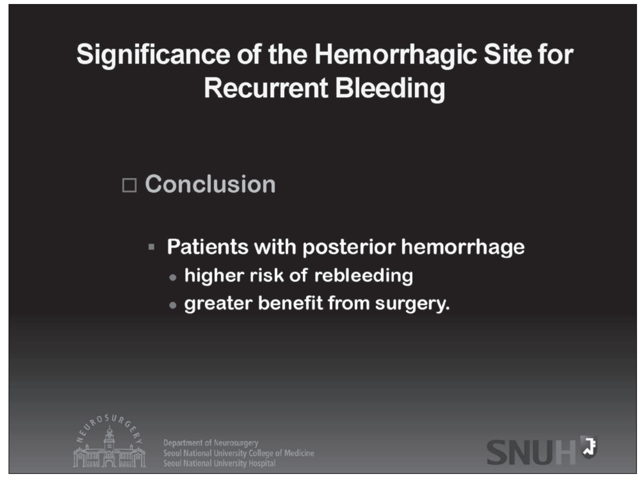

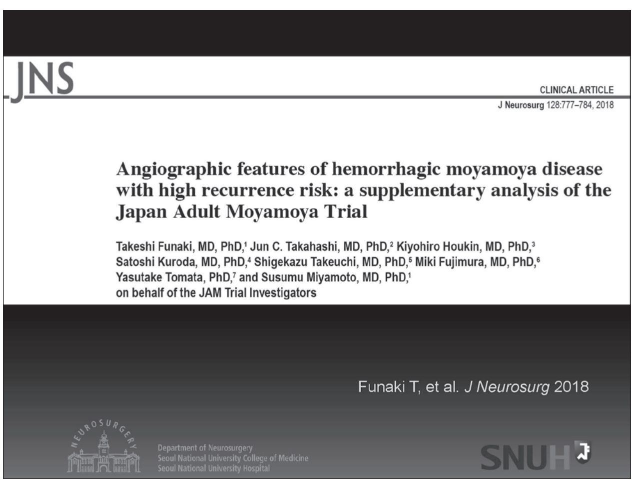

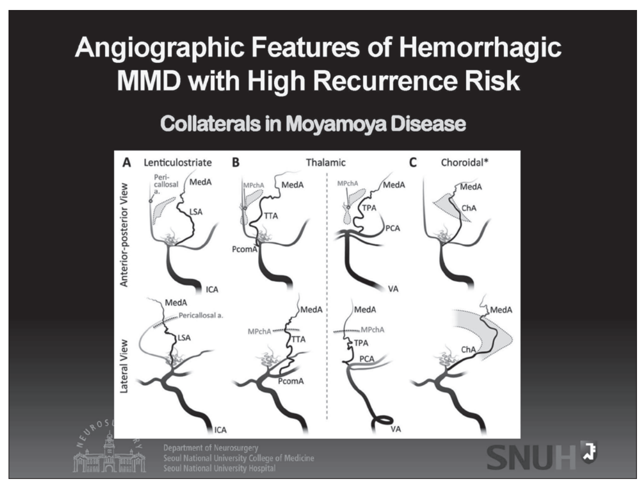

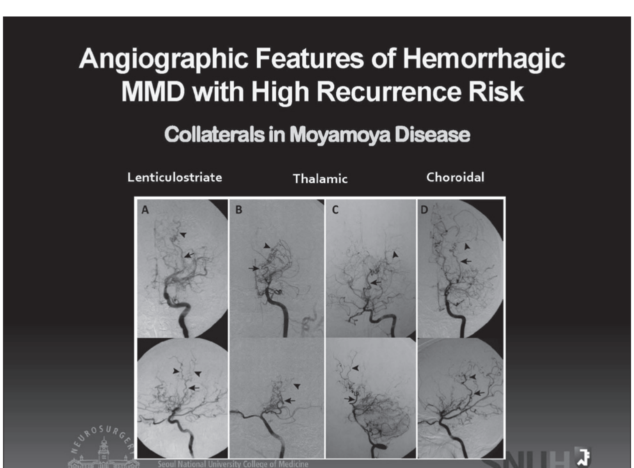

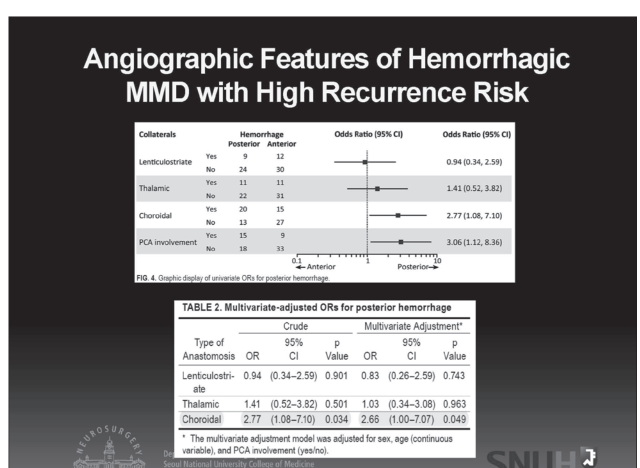

Collaterals		Hemorrhage Posterior	Hemorrhage Anterior	Odds Ratio (95% CI)
Lenticulostriate	Yes	9	12	0.94 (0.34, 2.59)
	No	24	30	
Thalamic	Yes	11	11	1.41 (0.52, 3.82)
	No	22	31	
Choroidal	Yes	20	15	2.77 (1.08, 7.10)
	No	13	27	
PCA involvement	Yes	15	9	3.06 (1.12, 8.36)
	No	18	33	

FIG. 4. Graphic display of univariate ORs for posterior hemorrhage.

TABLE 2. Multivariate-adjusted ORs for posterior hemorrhage

Type of Anastomosis	Crude OR	Crude 95% CI	Crude p Value	Multivariate Adjustment* OR	Multivariate Adjustment* 95% CI	Multivariate Adjustment* p Value
Lenticulostriate	0.94	(0.34–2.59)	0.901	0.83	(0.26–2.59)	0.743
Thalamic	1.41	(0.52–3.82)	0.501	1.03	(0.34–3.08)	0.963
Choroidal	2.77	(1.08–7.10)	0.034	2.66	(1.00–7.07)	0.049

* The multivariate adjustment model was adjusted for sex, age (continuous variable), and PCA involvement (yes/no).

Angiographic Features of Hemorrhagic MMD with High Recurrence Risk

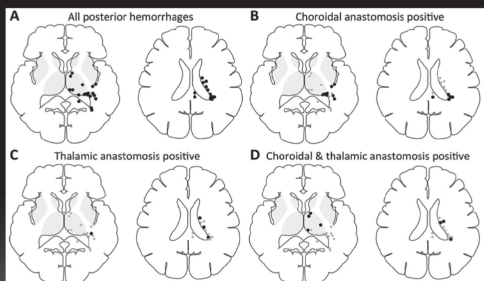

FIG. 5. Topographical analysis showing the relationship between bleeding points (*gray dots*) and collaterals. *Black dots* represent each collateral detected. For ease of recognition, all bleeding points are presented as on the left hemisphere. *Gray shading* indicates the basal ganglia and thalamus.

Grey dots : bleeding points
Black dots : collateral

Angiographic Features of Hemorrhagic MMD with High Recurrence Risk

- Choroidal anastomosis and PCA involvement
  - Characteristics of Posterior hemorrhage
- Choroidal anastomosis
  - Potential source of posterior hemorrhage at high risk of rebleeding

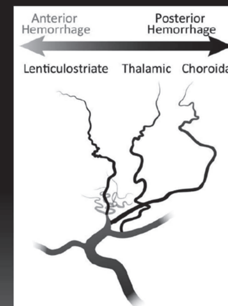

FIG. 6. Schematic illustration showing the conceptual relationship between collaterals and hemorrhage locations.

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JNS
CLINICAL ARTICLE
J Neurosurg 130:525–530, 2019

High rebleeding risk associated with choroidal collateral vessels in hemorrhagic moyamoya disease: analysis of a nonsurgical cohort in the Japan Adult Moyamoya Trial

Takeshi Funaki, MD, PhD,[1] Jun C. Takahashi, MD, PhD,[2] Kiyohiro Houkin, MD, PhD,[3] Satoshi Kuroda, MD, PhD,[4] Shigekazu Takeuchi, MD, PhD,[5] Miki Fujimura, MD, PhD,[6] Yasutake Tomata, PhD,[7] and Susumu Miyamoto, MD, PhD,[1] on behalf of the JAM Trial Investigators

Funaki T, et al. *Stroke* 2018

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High Rebleeding Risk Associated with Choroidal Collateral Vessels in Hemorrhagic MMD

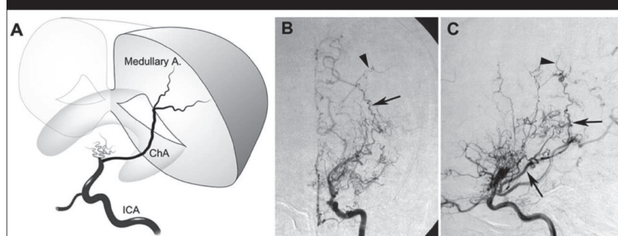

FIG. 1. Illustration (A) and representative angiograms, anteroposterior (B) and lateral (C) views, showing choroidal anastomosis. Dilated and extended anterior or posterior choroidal artery (*arrows*) connects to the medullary artery (*arrowheads*). Although the illustration represents choroidal anastomosis from the anterior choroidal artery, together the anterior choroidal artery from the internal carotid artery and the lateral posterior choroidal arteries from the PCA can serve as the origin of such an anastomosis. A. = artery; ChA = choroidal artery; ICA = internal carotid artery. Illustration copyright Takeshi Funaki. Published with permission.

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High Rebleeding Risk Associated with Choroidal Collateral Vessels in Hemorrhagic MMD

- 37 patients – nonsurgical arm
  - Choroidal collaterals (+) : 21

TABLE 1. Baseline variables in 37 moyamoya patients in the nonsurgical group

Variable	Choroidal Anastomosis Positive	Choroidal Anastomosis Negative	p Value
No. of patients	21	16	
Mean age in yrs ± SD	39.9 ± 13.4	43.4 ± 11.1	0.405
Female	17 (81.0)	10 (62.5)	0.274
Hypertension	4 (19.0)	3 (18.8)	1.000
Diabetes mellitus	1 (4.8)	1 (6.3)	1.000
Hyperlipidemia	1 (4.8)	1 (6.3)	1.000
Hx of ischemic event	7 (33.3)	3 (18.8)	0.461
Hx of hemorrhagic stroke	2 (9.5)	2 (12.5)	1.000
PCA involvement	9 (42.9)	3 (18.8)	0.121

Hx = history.
Values expressed as number (%) of patients, unless otherwise indicated.

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High Rebleeding Risk Associated with Choroidal Collateral Vessels in Hemorrhagic MMD

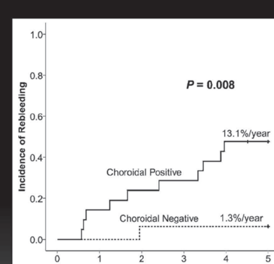

FIG. 2. Kaplan-Meier curve for rebleeding (37 patients).

- Rebleeding
  - 11 of 37 cases
  - 7.2% per year
- Rebleeding rate
  - Choroidal collateral (+)
    - 5-year → 47.6%
  - Choroidal collateral (-)
    - 5-year → 6.5%

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High Rebleeding Risk Associated with Choroidal Collateral Vessels in Hemorrhagic MMD

- Longitudinal cohort analysis from JAM trials
- Choroidal collaterals in hemorrhagic MMD
  - Bleeding source with high risk for hemorrhagic recurrence
  - Predictor of rebleeding

JNS
CLINICAL ARTICLE
J Neurosurg 130:1453–1459, 2019

Intrinsic development of choroidal and thalamic collaterals in hemorrhagic-onset moyamoya disease: case-control study of the Japan Adult Moyamoya Trial

Miki Fujimura, MD, PhD,[1] Takeshi Funaki, MD, PhD,[2] Kiyohiro Houkin, MD, PhD,[3]
Jun C. Takahashi, MD, PhD,[4] Satoshi Kuroda, MD, PhD,[5] Yasutake Tomata, PhD,[6]
Teiji Tominaga, MD, PhD,[1] Susumu Miyamoto, MD, PhD,[2] on behalf of the Japan Adult Moyamoya Trial Investigators

Fujimura M, et al. *J Neurosurg* 2018

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Intrinsic Development of Choroidal and Thalamic Collaterals in Hemorrhagic-onset MMD

- To identify the angiographic features
  - Hemorrhagic-onset versus Ischemic-onset

TABLE 1. Angiographic scoring of lenticulostriate, thalamic, and choroidal anastomosis in MMD

Collateral	Grade 0	Grade 1	Grade 2
Lenticulostriate	No dilation, no extension	Dilation/extension below the level of the pericallosal artery	Dilation/extension beyond the level of the pericallosal artery
Thalamic (perforator)	No dilation, no extension	Dilation/extension below the level of the MPChA	Dilation/extension beyond the level of the MPChA
Choroidal	No dilation, no extension	Dilation/extension below the level of the lateral ventricle	Dilation/extension beyond the level of lateral ventricle

MPChA = medial posterior choroidal artery.

Grade 2 : positive angiographic indicator of choroidal anastomosis

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Intrinsic Development of Choroidal and Thalamic Collaterals in Hemorrhagic-onset MMD

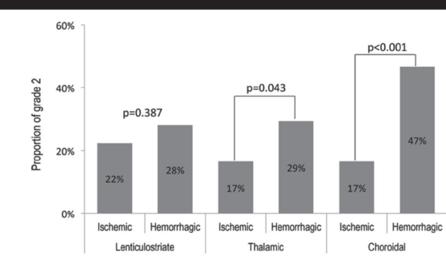

FIG. 2. The prevalence of high angiographic score (grade 2) according to each collateral pattern. Statistical significance was determined by chi-square test. Figure is available in color online only.

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Intrinsic Development of Choroidal and Thalamic Collaterals in Hemorrhagic-onset MMD

TABLE 3. Proportions (adjusted odds ratios) of grade 2 MMD in each collateral pattern

	Grade ≤1		Grade 2		Crude			Sex/Age Adjusted		
Collateral	No.	%	No.	%	OR	95% CI	p Value	OR	95% CI	p Value
Lenticulostriate anastomosis										
Ischemic	80	77.7%	23	22.3%	1.00	Reference		1.00	Reference	
Hemorrhagic	54	72.0%	21	28.0%	1.35	0.68–2.68	0.387	1.88	0.89–3.96	0.097
Thalamic anastomosis										
Ischemic	86	83.5%	17	16.5%	1.00	Reference		1.00	Reference	
Hemorrhagic	53	70.7%	22	29.3%	2.10	1.02–4.31	0.043	2.18	1.03–4.60	0.041
Choroidal anastomosis										
Ischemic	86	83.5%	17	16.5%	1.00	Reference		1.00	Reference	
Hemorrhagic	40	53.3%	35	46.7%	4.43	2.22–8.83	<0.001	4.74	2.31–9.72	<0.001

Overall 178 hemispheres (n = 103 ischemic and 75 hemorrhagic).

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Intrinsic Development of Choroidal and Thalamic Collaterals in Hemorrhagic-onset MMD

- Involvement of the PCA

TABLE 4. Proportions (adjusted odds ratios) of PCA involvement

	PCA Involvement									
	Grade 0		Grade ≥1		Crude			Sex/Age Adjusted		
MMD Group	No.	%	No.	%	OR	95% CI	p Value	OR	95% CI	p Value
Ischemic	81	78.6%	22	21.4%	1.00	Reference		1.00	Reference	
Hemorrhagic	51	68.0%	24	32.0%	1.73	0.88–3.41	0.111	1.60	0.79–3.21	0.189

Overall 178 hemispheres (n = 103 ischemic and 75 hemorrhagic).

- Suzuki's angiographic staging

TABLE 5. Comparison of Suzuki angiographic stage between ischemic-onset and hemorrhagic-onset groups

	Suzuki Stage									
	Grade <4		Grade ≥4		Crude			Sex/Age Adjusted		
MMD Group	No.	%	No.	%	OR	95% CI	p Value	OR	95% CI	p Value
Ischemic	49	47.6%	54	52.4%	1.00	Reference		1.00	Reference	
Hemorrhagic	24	32.0%	51	68.0%	1.93	1.04–3.59	0.038	1.95	1.02–3.74	0.045

Overall 178 hemispheres (n = 103 ischemic and 75 hemorrhagic).

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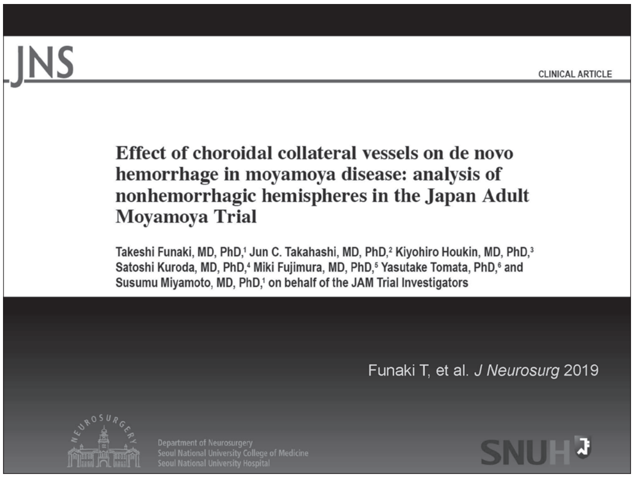


JNS CLINICAL ARTICLE

Effect of choroidal collateral vessels on de novo hemorrhage in moyamoya disease: analysis of nonhemorrhagic hemispheres in the Japan Adult Moyamoya Trial

Takeshi Funaki, MD, PhD,[1] Jun C. Takahashi, MD, PhD,[2] Kiyohiro Houkin, MD, PhD,[3] Satoshi Kuroda, MD, PhD,[4] Miki Fujimura, MD, PhD,[5] Yasutake Tomata, PhD,[6] and Susumu Miyamoto, MD, PhD,[1] on behalf of the JAM Trial Investigators

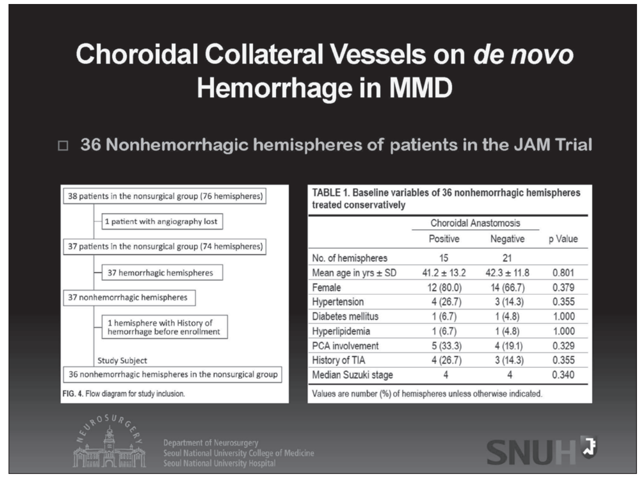
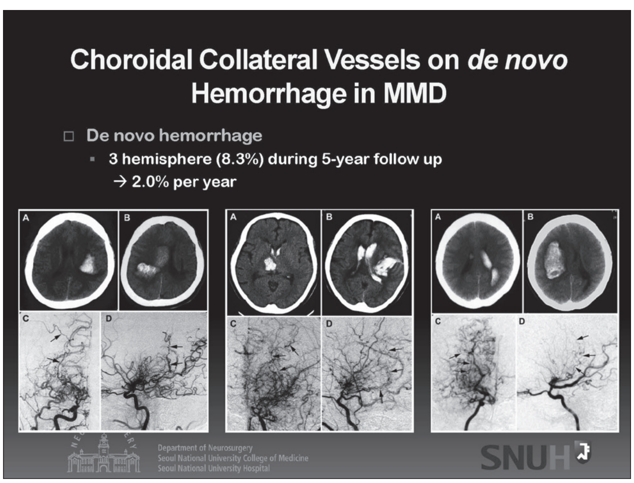
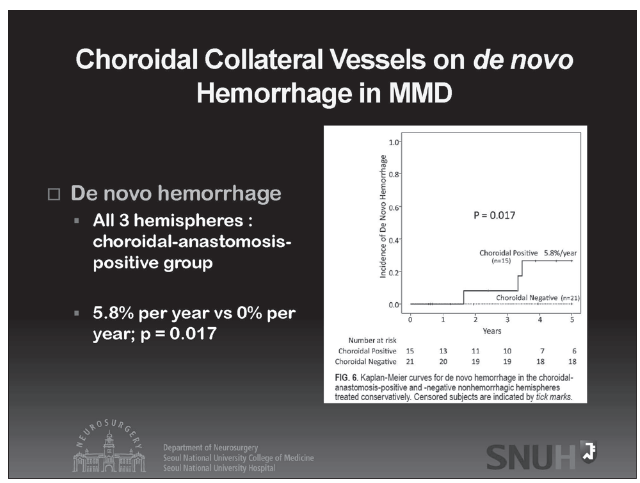


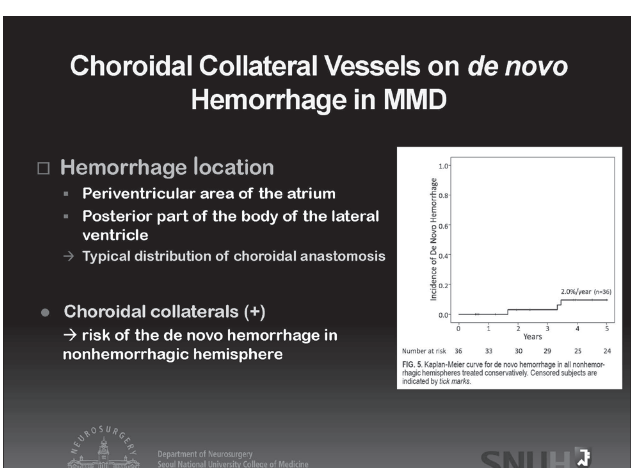
TABLE 1. Baseline variables of 36 nonhemorrhagic hemispheres treated conservatively

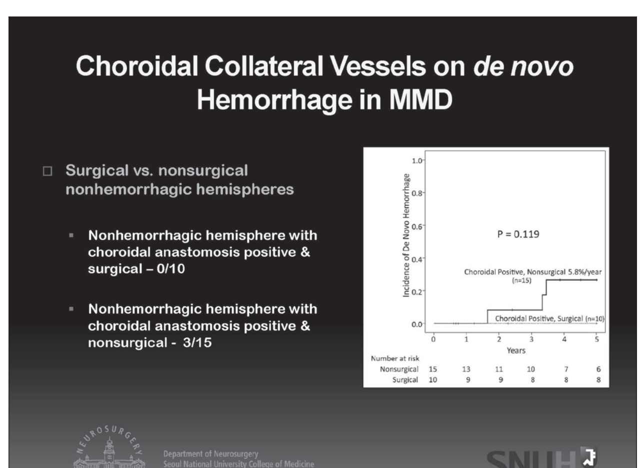
	Choroidal Anastomosis		
	Positive	Negative	p Value
No. of hemispheres	15	21	
Mean age in yrs ± SD	41.2 ± 13.2	42.3 ± 11.8	0.801
Female	12 (80.0)	14 (66.7)	0.379
Hypertension	4 (26.7)	3 (14.3)	0.355
Diabetes mellitus	1 (6.7)	1 (4.8)	1.000
Hyperlipidemia	1 (6.7)	1 (4.8)	1.000
PCA involvement	5 (33.3)	4 (19.1)	0.329
History of TIA	4 (26.7)	3 (14.3)	0.355
Median Suzuki stage	4	4	0.340

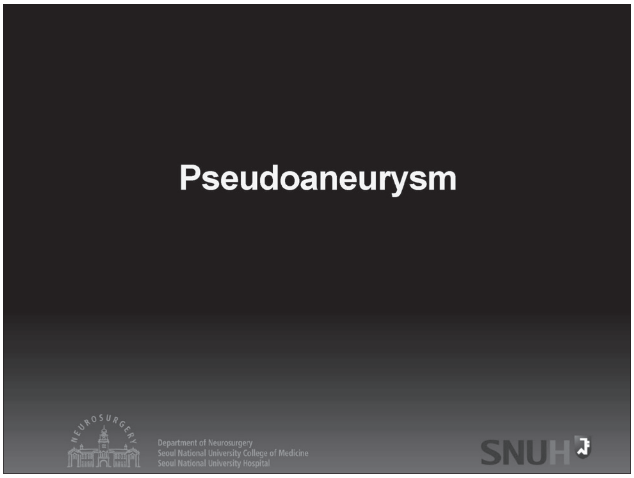
Values are number (%) of hemispheres unless otherwise indicated.










Pseudoaneurysm
Department of Neurosurgery
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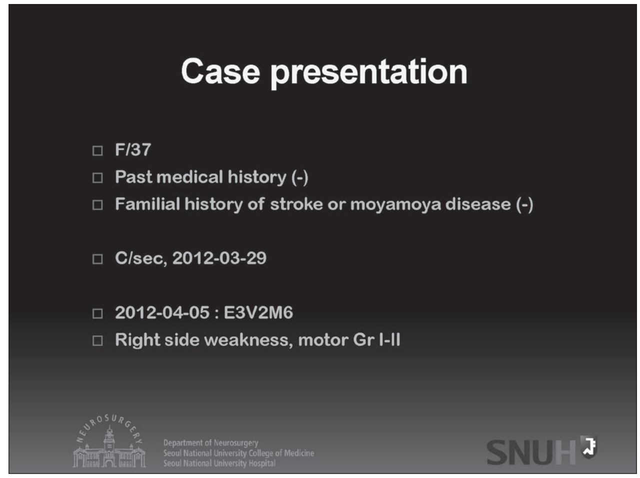
Seoul National University Hospital


Case presentation
F/37
Past medical history (-)
Familial history of stroke or moyamoya disease (-)
C/sec, 2012-03-29
2012-04-05 : E3V2M6
Right side weakness, motor Gr I-II
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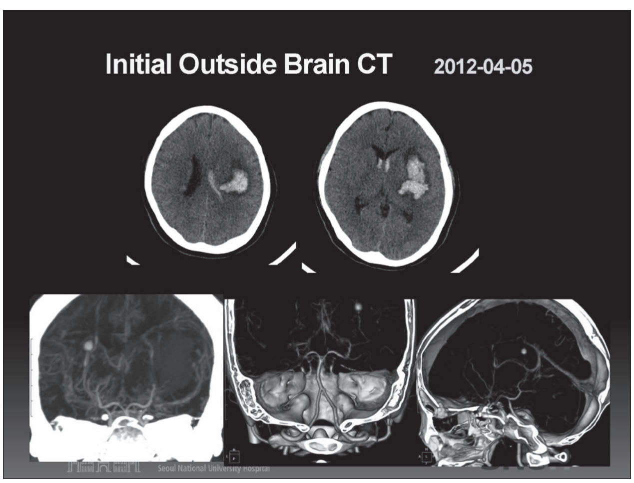
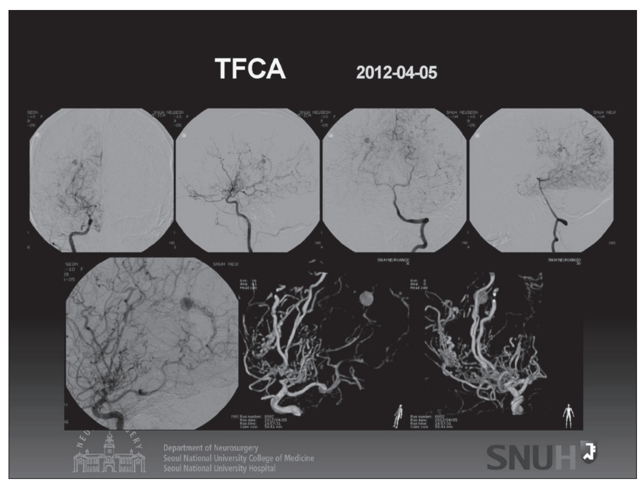
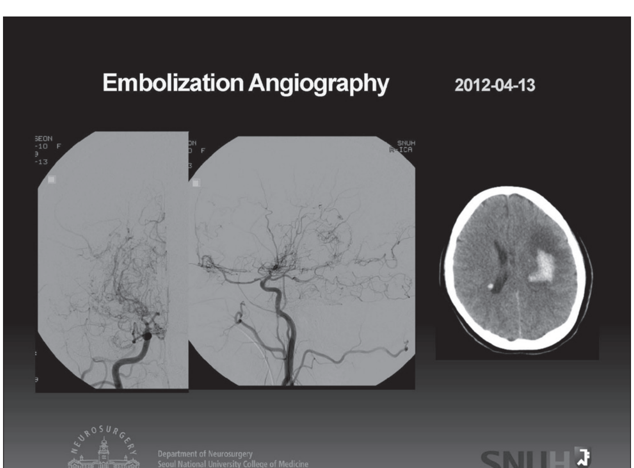
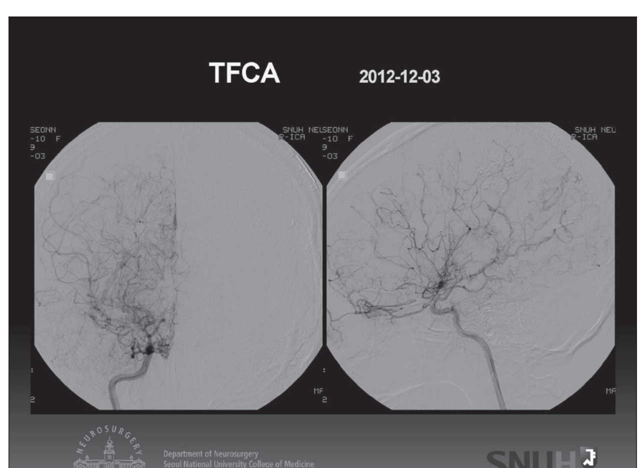
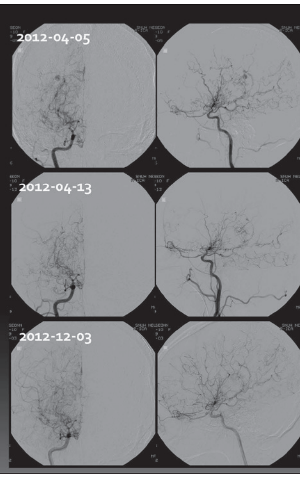

Initial Outside Brain CT
2012-04-05


TFCA
2012-04-05


Embolization Angiography
2012-04-13


TFCA
2012-12-03

Change in TFCA



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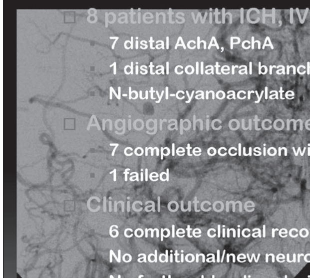
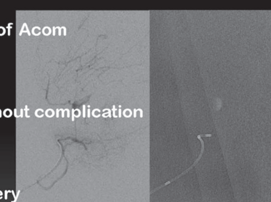
CLINICAL REPORT

Neurosurgery 2009

ENDOVASCULAR TREATMENT OF RUPTURED ANEURYSMS OR PSEUDOANEURYSMS ON THE COLLATERAL VESSELS IN PATIENTS WITH MOYAMOYA DISEASE

Kim SH, et al. *Neurosurgery* 2009





- 8 patients with ICH, IVH
  - 7 distal AchA, PchA
  - 1 distal collateral branch of Acom
  - N-butyl-cyanoacrylate
- Angiographic outcome
  - 7 complete occlusion without complication
  - 1 failed
- Clinical outcome
  - 6 complete clinical recovery
  - No additional/new neurologic deficit / infarction
  - No further bleeding during 24 months (11-60) follow-up

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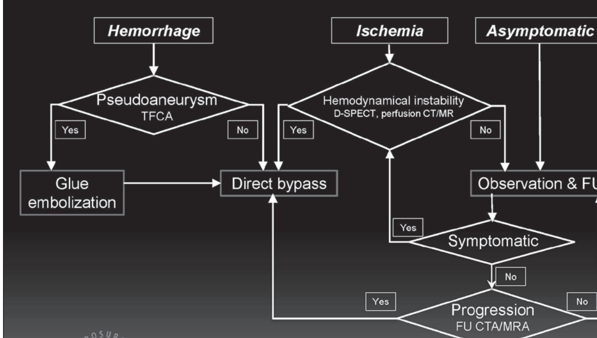
Surgical Indications *SNUH*

- Symptomatic
  - Repeated hemorrhage
  - Asymptomatic but progression on MRA, CTA, TFCA
- Functionally independent: KPS ≥ 70
- No acute infarction/hemorrhage within 4 weeks
- Hemodynamically unstable
  - D-SPECT with perfusion CT or perfusion MR ASL

SNUH

My Bypass Policy for Adult MMD *SNUH*



SNUH

Conclusion

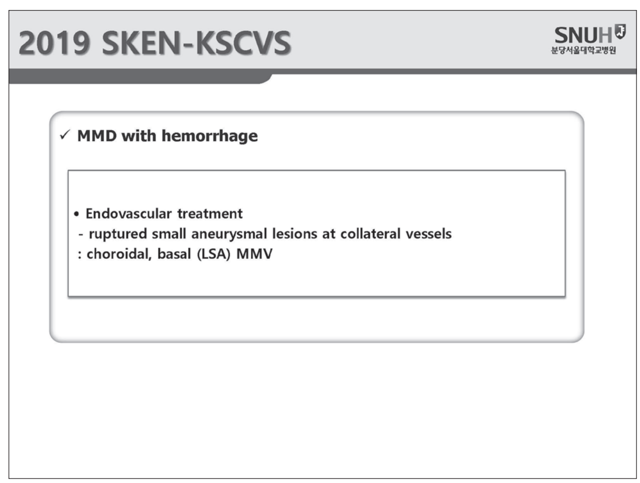
- Direct revascularization is an effective treatment for hemorrhagic adult MMD to prevent further hemorrhage on the affected hemisphere.

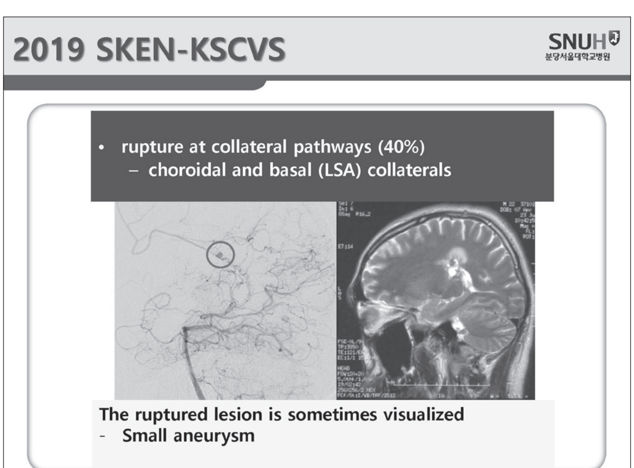
SNUH

Endovascular treatment for targeted embolization in Moyamoya disease with hemorrhagic manifestation

반승필

서울대





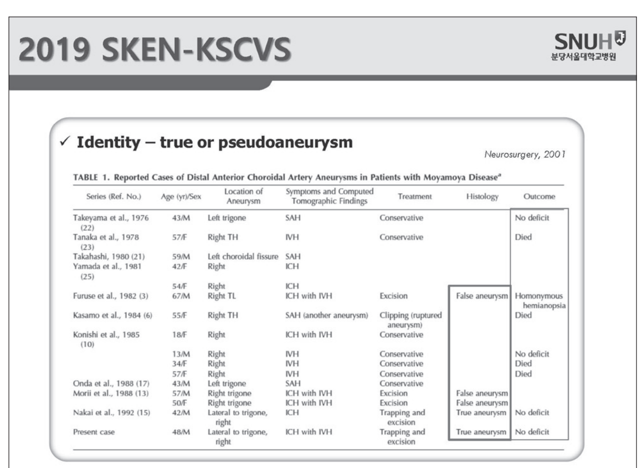
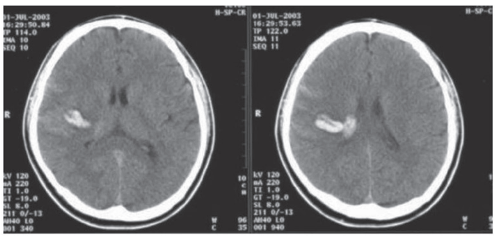


TABLE 1. Reported Cases of Distal Anterior Choroidal Artery Aneurysms in Patients with Moyamoya Disease[a]

Series (Ref. No.)	Age (yr)/Sex	Location of Aneurysm	Symptoms and Computed Tomographic Findings	Treatment	Histology	Outcome
Takeyama et al., 1976 (22)	43/M	Left trigone	SAH	Conservative		No deficit
Tanaka et al., 1978 (23)	57/F	Right TH	IVH	Conservative		Died
Takahashi, 1980 (21)	59/M	Left choroidal fissure	SAH			
Yamada et al., 1981 (25)	42/F	Right	ICH			
	54/F	Right	ICH			
Furuse et al., 1982 (3)	67/M	Right TL	ICH with IVH	Excision	False aneurysm	Homonymous hemianopsia
Kasamo et al., 1984 (6)	55/F	Right TH	SAH (another aneurysm)	Clipping (ruptured aneurysm)		Died
Konishi et al., 1985 (10)	18/F	Right	ICH with IVH	Conservative		
	13/M	Right	IVH	Conservative		No deficit
	34/F	Right	IVH	Conservative		Died
	57/F	Right	IVH	Conservative		Died
Onda et al., 1988 (17)	43/M	Left trigone	SAH	Conservative		
Morii et al., 1988 (13)	57/M	Right trigone	ICH with IVH	Excision	False aneurysm	
	50/F	Right trigone	ICH with IVH	Excision	False aneurysm	
Nakai et al., 1992 (15)	42/M	Lateral to trigone, right	ICH	Trapping and excision	True aneurysm	No deficit
Present case	48/M	Lateral to trigone, right	ICH with IVH	Trapping and excision	True aneurysm	No deficit

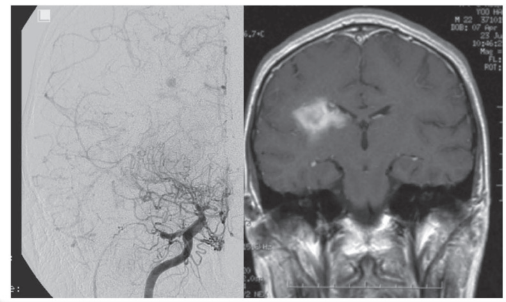
2019 SKEN-KSCVS

✓ **M/23, Left hemiparesis Gr. 4**



2019 SKEN-KSCVS

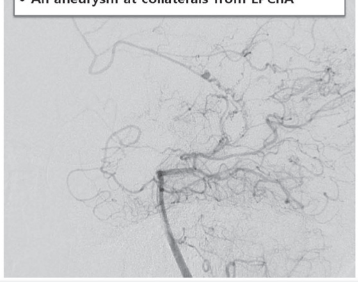
✓ **M/23, Left hemiparesis Gr. 4**



2019 SKEN-KSCVS

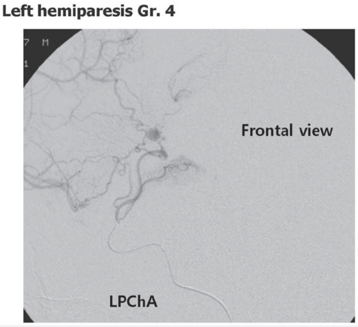
✓ **M/23, Left hemiparesis Gr. 4**

- An aneurysm at collaterals from LPChA



2019 SKEN-KSCVS

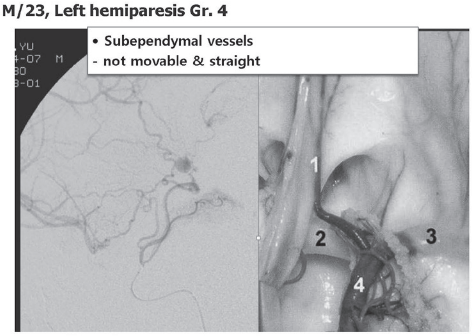
✓ **M/23, Left hemiparesis Gr. 4**



2019 SKEN-KSCVS

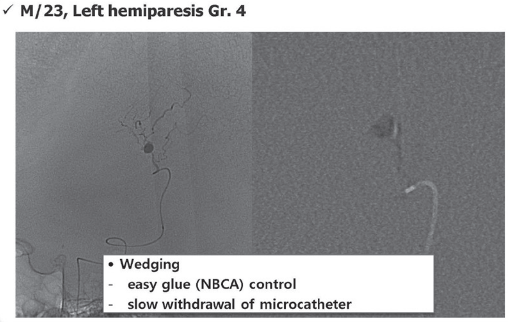
✓ **M/23, Left hemiparesis Gr. 4**

- Subependymal vessels
- not movable & straight

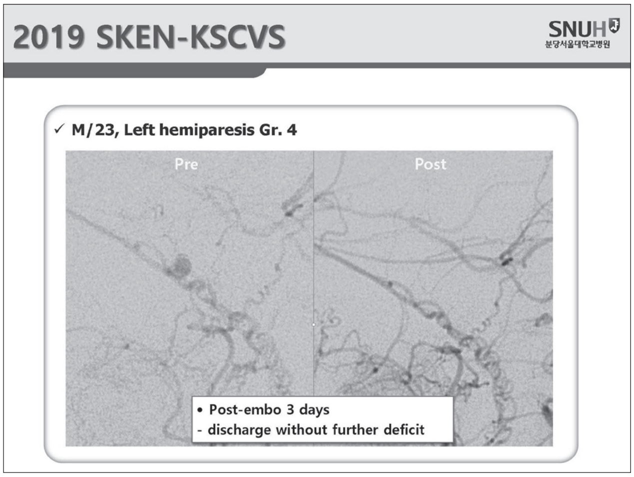


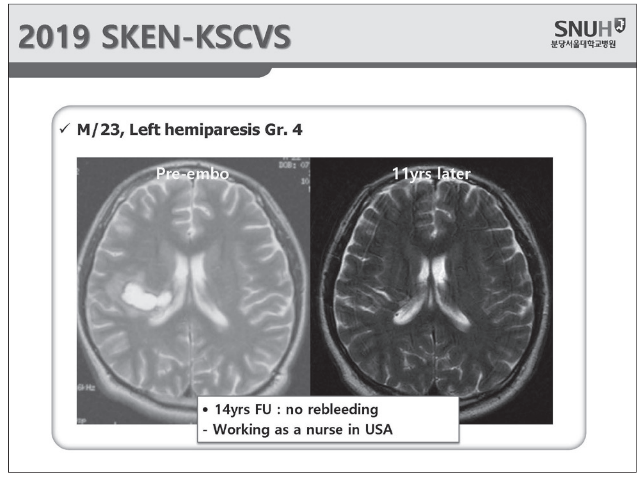
2019 SKEN-KSCVS

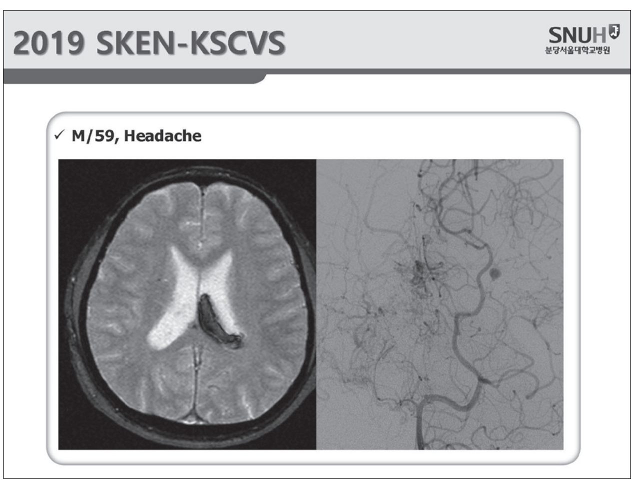
✓ **M/23, Left hemiparesis Gr. 4**

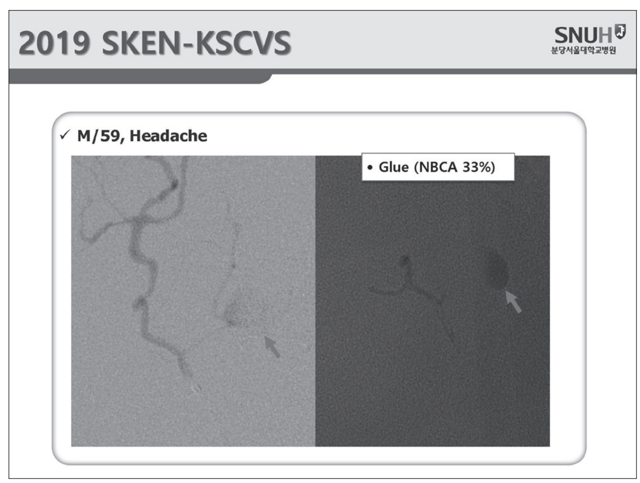


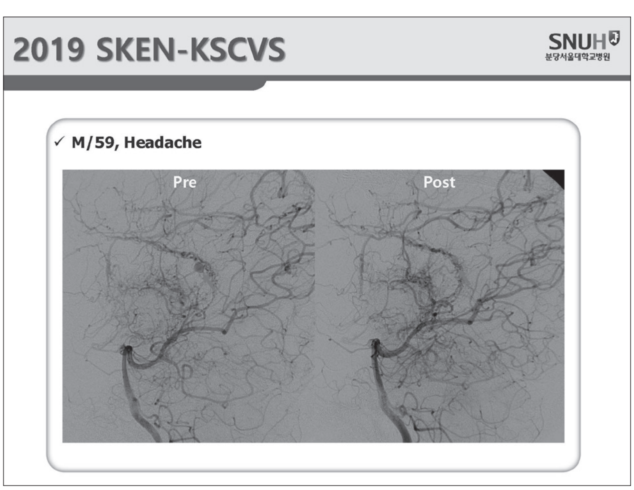
- Wedging
- easy glue (NBCA) control
- slow withdrawal of microcatheter

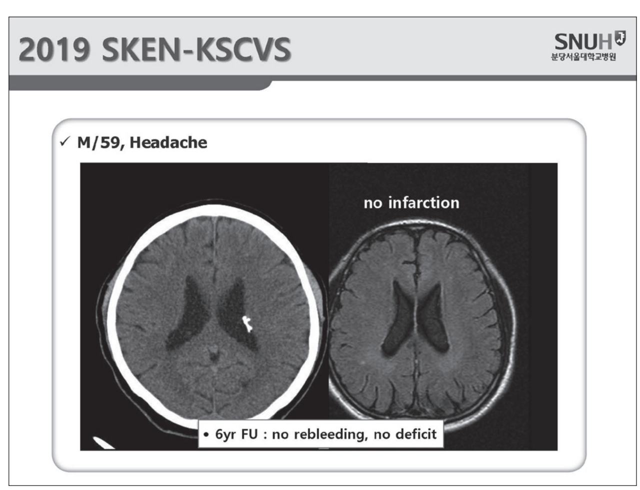

2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ M/23, Left hemiparesis Gr. 4
Pre
Post
• Post-embo 3 days
- discharge without further deficit


2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ M/23, Left hemiparesis Gr. 4
Pre-embo
11yrs later
• 14yrs FU : no rebleeding
- Working as a nurse in USA


2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ M/59, Headache

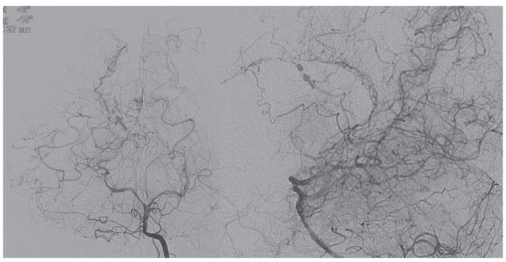

2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ M/59, Headache
• Glue (NBCA 33%)


2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ M/59, Headache
Pre
Post


2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ M/59, Headache
no infarction
• 6yr FU : no rebleeding, no deficit

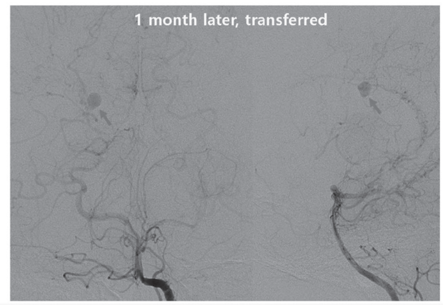
2019 SKEN-KSCVS

✓ F/62, Headache / IVH_EVD and observation



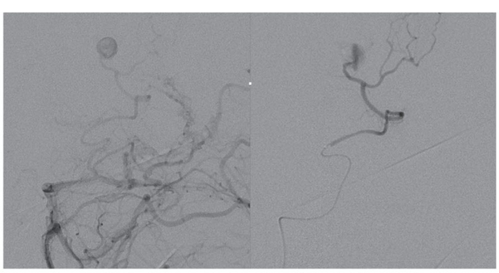
2019 SKEN-KSCVS

✓ F/62, Headache / IVH_EVD and observation



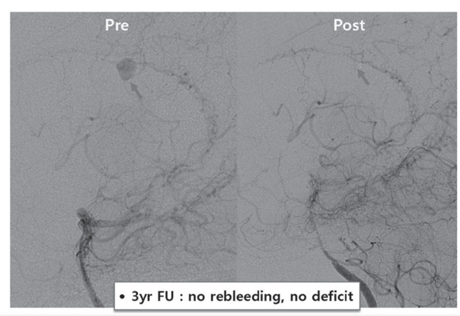
2019 SKEN-KSCVS

✓ F/62, Headache / IVH_EVD and observation



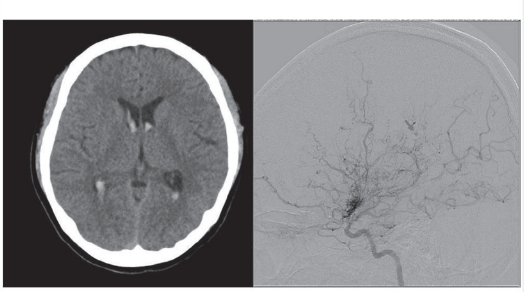
2019 SKEN-KSCVS

✓ F/62, Headache / IVH_EVD and observation



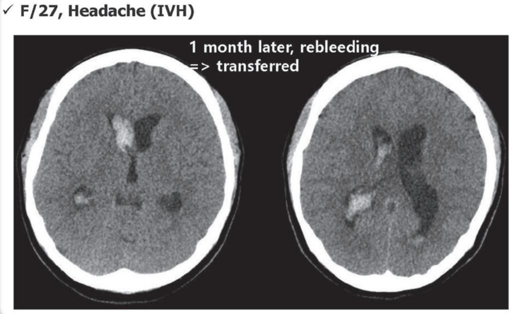
2019 SKEN-KSCVS

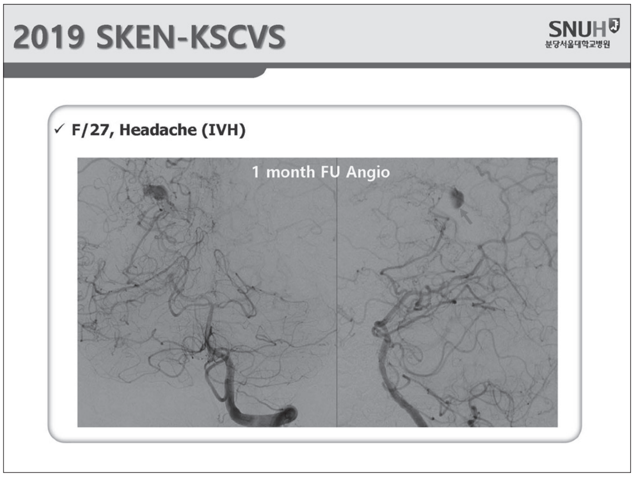
✓ F/27, Headache -> IVH & observation

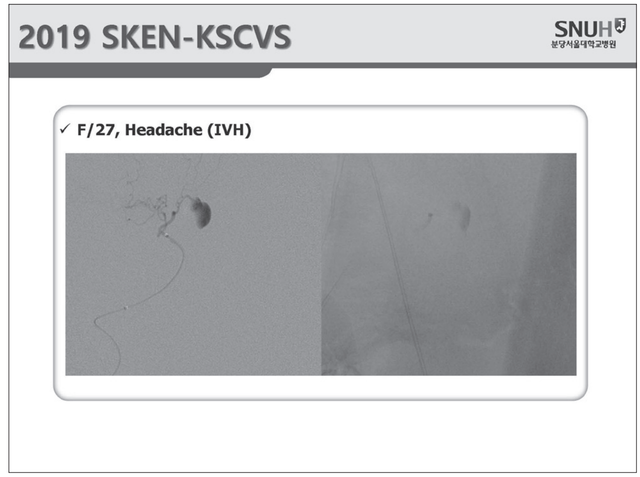


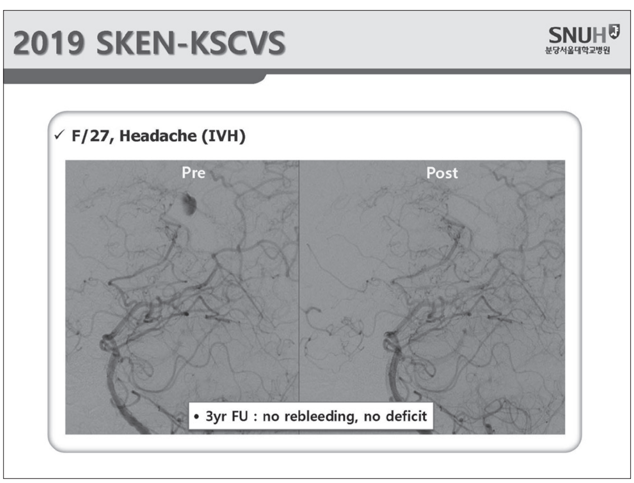
2019 SKEN-KSCVS

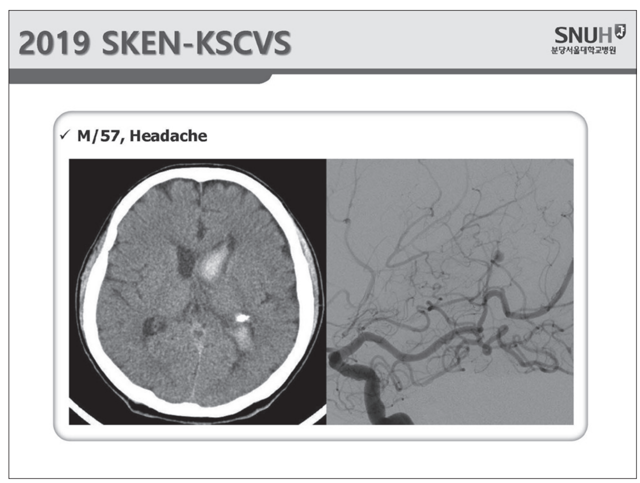
✓ F/27, Headache (IVH)

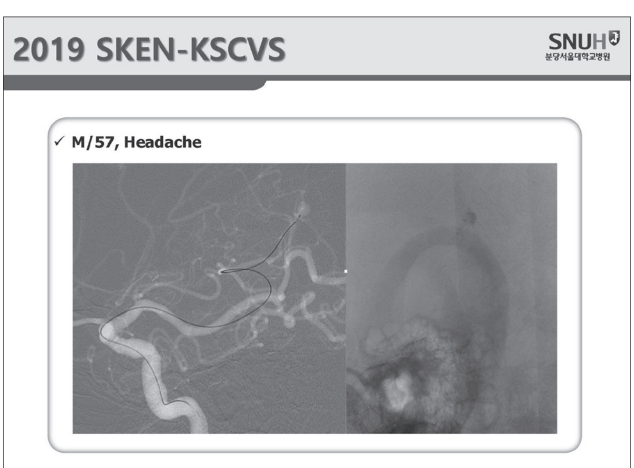


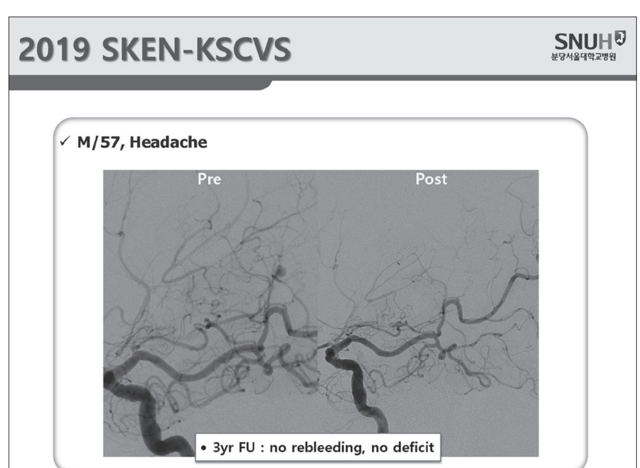

2019 SKEN-KSCVS
SNUH
✓ F/27, Headache (IVH)
1 month FU Angio


2019 SKEN-KSCVS
SNUH
✓ F/27, Headache (IVH)


2019 SKEN-KSCVS
SNUH
✓ F/27, Headache (IVH)
Pre
Post
• 3yr FU : no rebleeding, no deficit

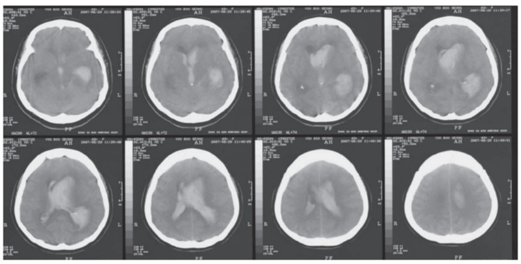

2019 SKEN-KSCVS
SNUH
✓ M/57, Headache


2019 SKEN-KSCVS
SNUH
✓ M/57, Headache


2019 SKEN-KSCVS
SNUH
✓ M/57, Headache
Pre
Post
• 3yr FU : no rebleeding, no deficit

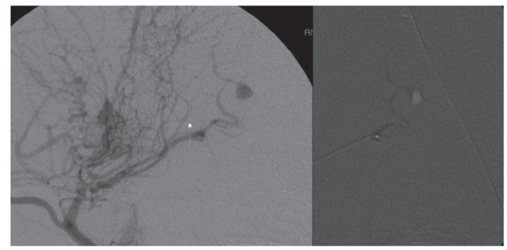
2019 SKEN-KSCVS

✓ **F/43, Coma**



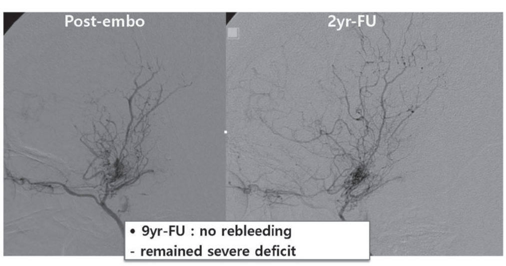
2019 SKEN-KSCVS

✓ **F/43, Coma**



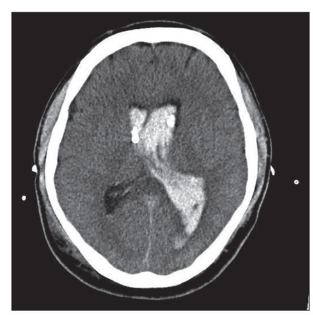
2019 SKEN-KSCVS

✓ **F/43, Coma**



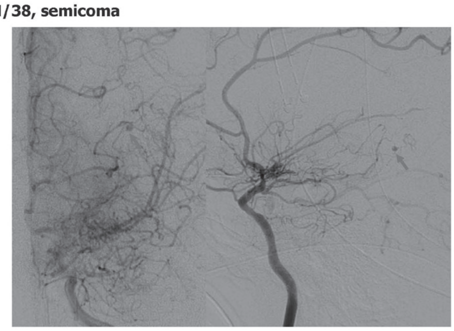
2019 SKEN-KSCVS

✓ **M/38, semicoma**



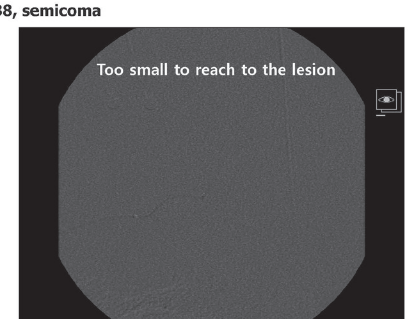
2019 SKEN-KSCVS

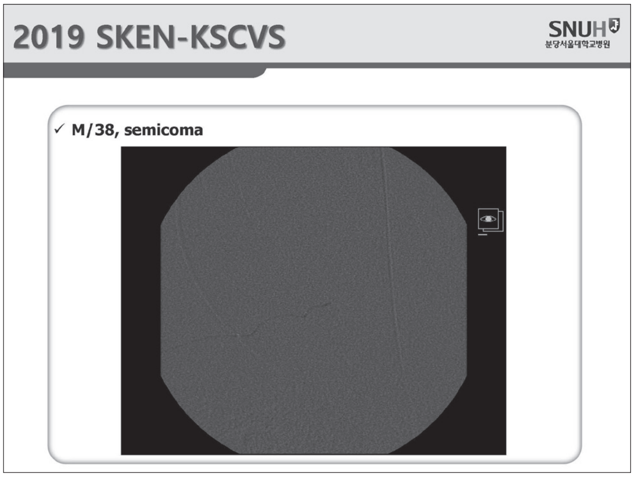
✓ **M/38, semicoma**

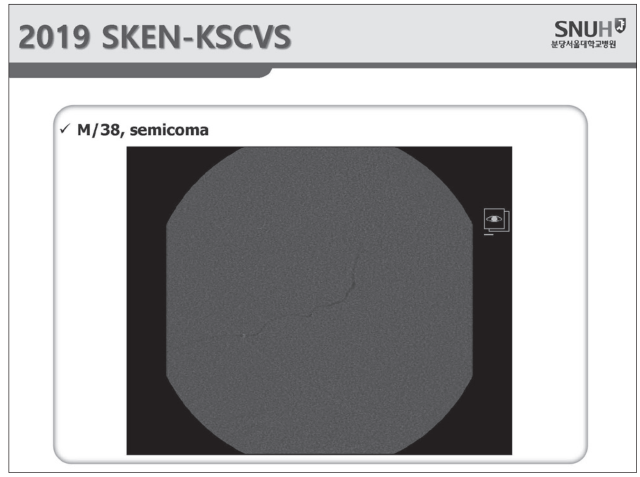


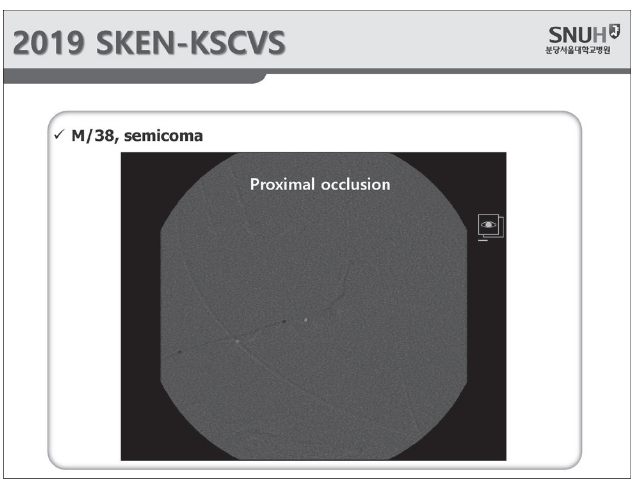
2019 SKEN-KSCVS

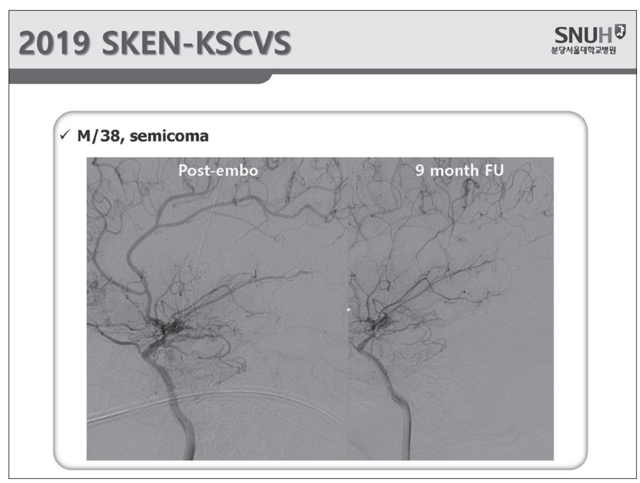
✓ **M/38, semicoma**

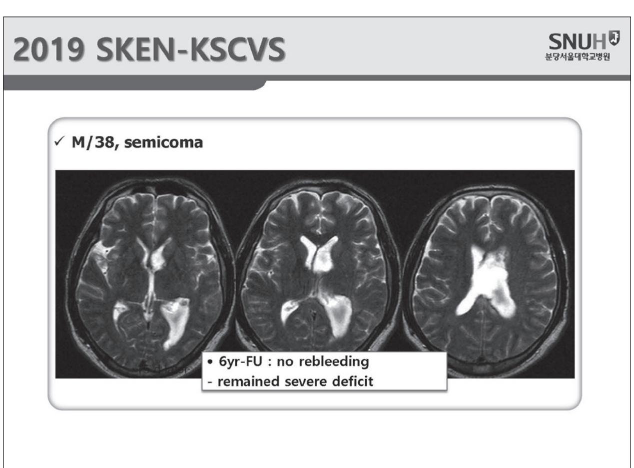


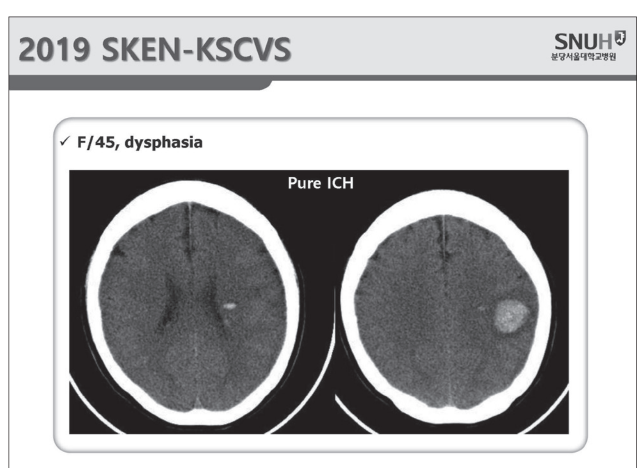

2019 SKEN-KSCVS
SNUH
✓ M/38, semicoma


2019 SKEN-KSCVS
SNUH
✓ M/38, semicoma


2019 SKEN-KSCVS
SNUH
✓ M/38, semicoma
Proximal occlusion


2019 SKEN-KSCVS
SNUH
✓ M/38, semicoma
Post-embo
9 month FU

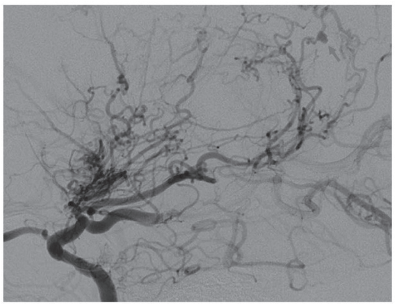

2019 SKEN-KSCVS
SNUH
✓ M/38, semicoma
• 6yr-FU : no rebleeding
- remained severe deficit


2019 SKEN-KSCVS
SNUH
✓ F/45, dysphasia
Pure ICH

2019 SKEN-KSCVS

SNUH 분당서울대학교병원

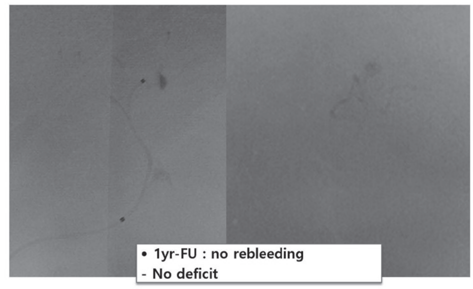
✓ **F/45, dysphasia**



2019 SKEN-KSCVS

SNUH 분당서울대학교병원

✓ **F/45, dysphasia**

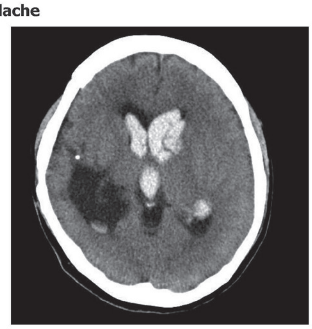


- 1yr-FU : no rebleeding
- No deficit

2019 SKEN-KSCVS

SNUH 분당서울대학교병원

✓ **F/44, headache**



2019 SKEN-KSCVS

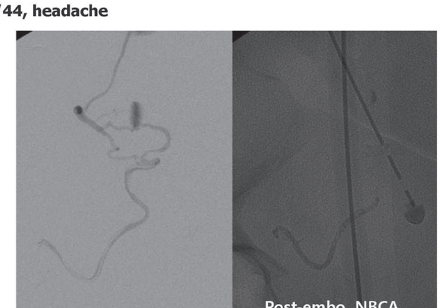
SNUH 분당서울대학교병원

✓ **F/44, headache**

2019 SKEN-KSCVS

SNUH 분당서울대학교병원

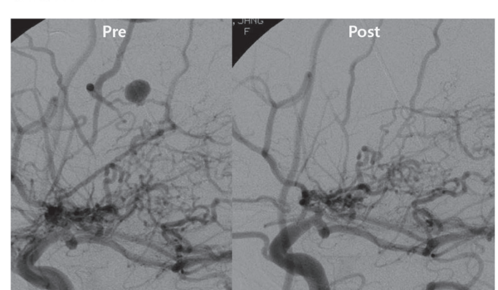
✓ **F/44, headache**

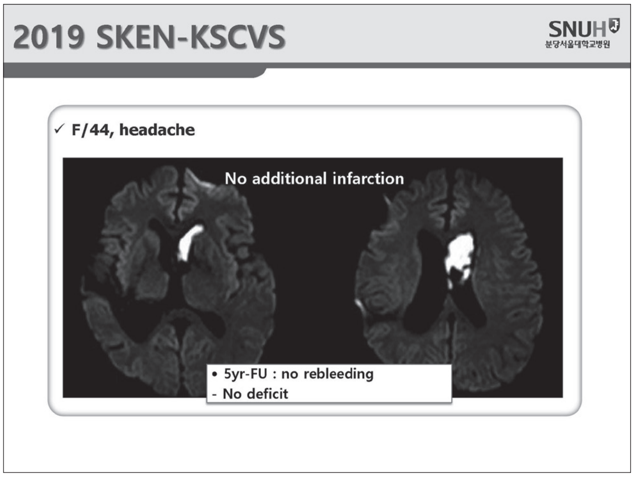


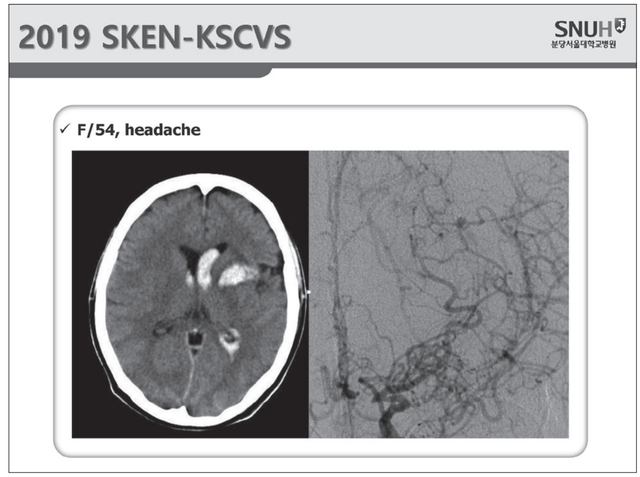
2019 SKEN-KSCVS

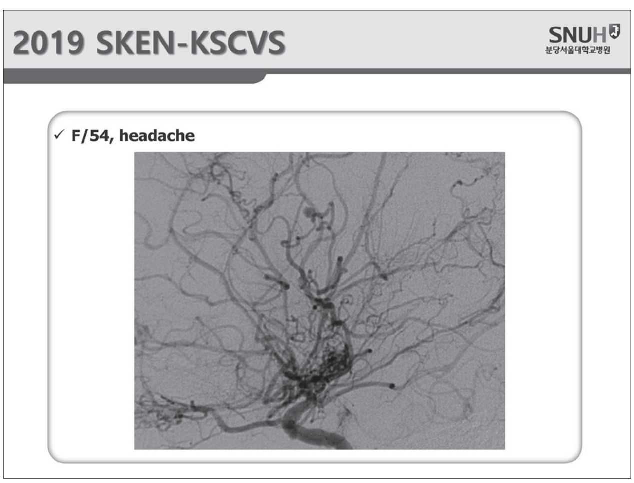
SNUH 분당서울대학교병원

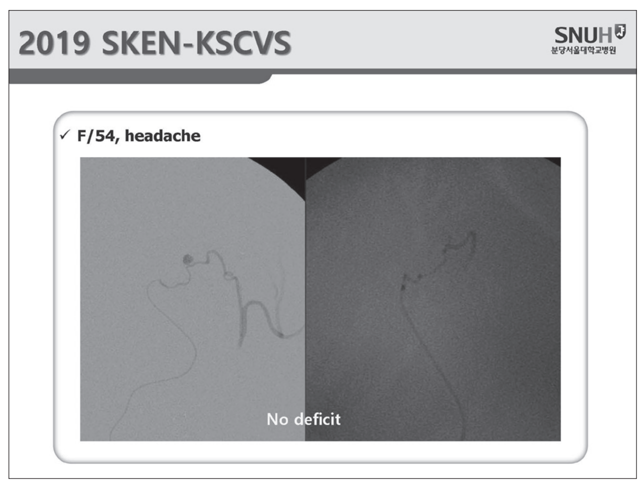
✓ **F/44, headache**

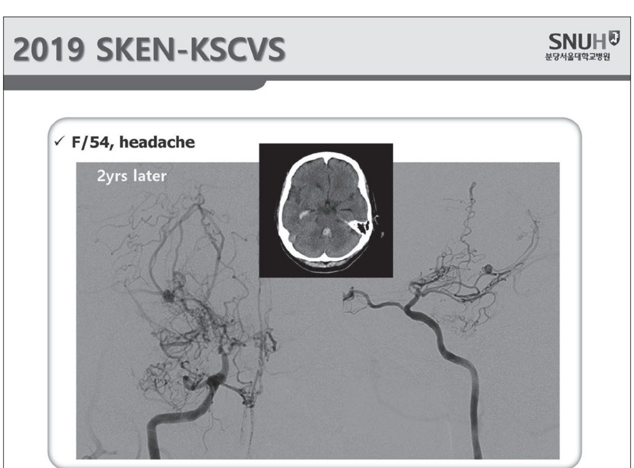


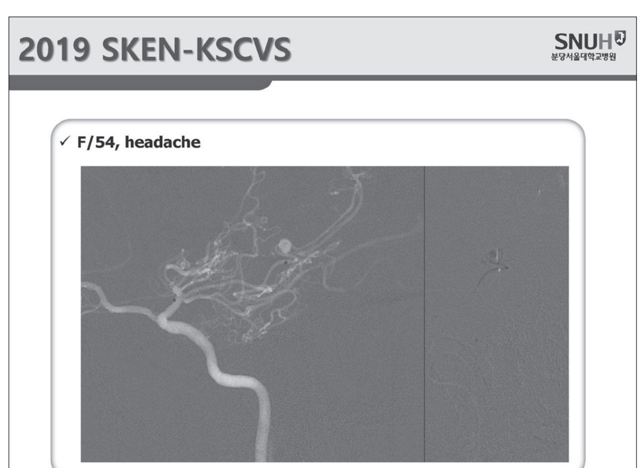

2019 SKEN-KSCVS
✓ F/44, headache
No additional infarction
• 5yr-FU : no rebleeding
- No deficit

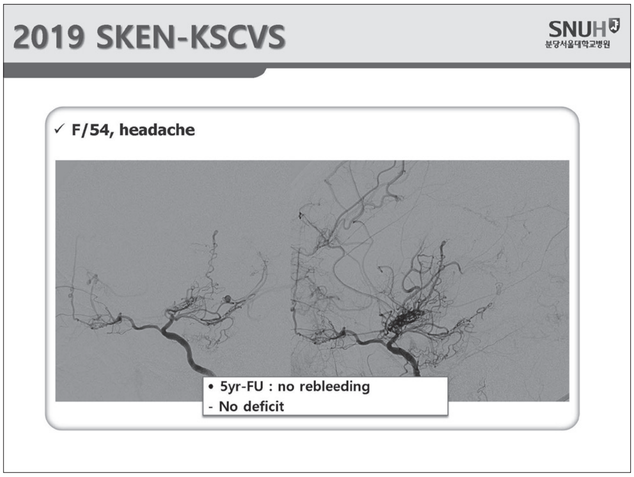

2019 SKEN-KSCVS
✓ F/54, headache

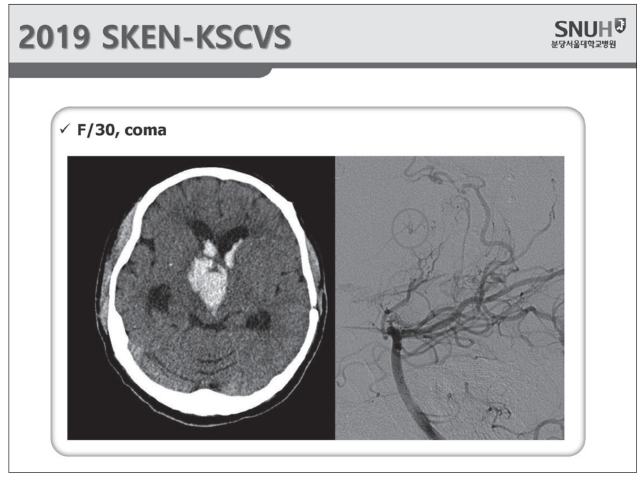

2019 SKEN-KSCVS
✓ F/54, headache

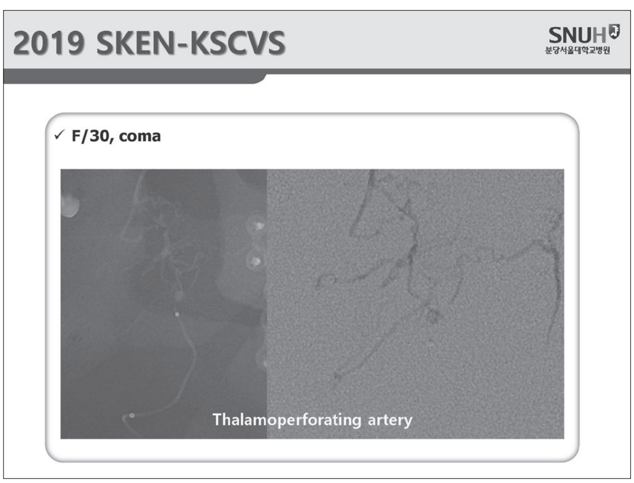

2019 SKEN-KSCVS
✓ F/54, headache
No deficit

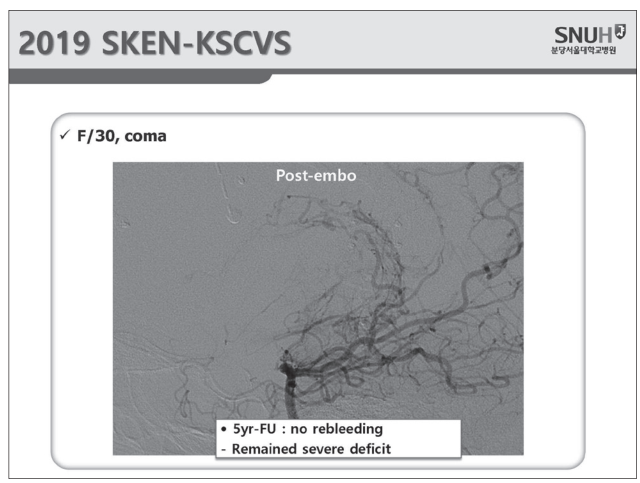

2019 SKEN-KSCVS
✓ F/54, headache
2yrs later

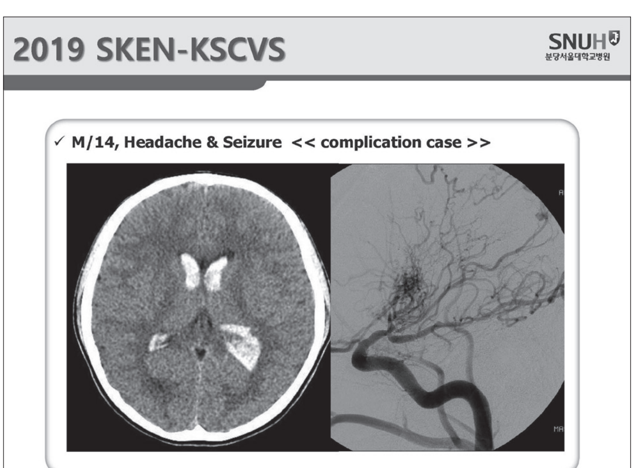

2019 SKEN-KSCVS
✓ F/54, headache

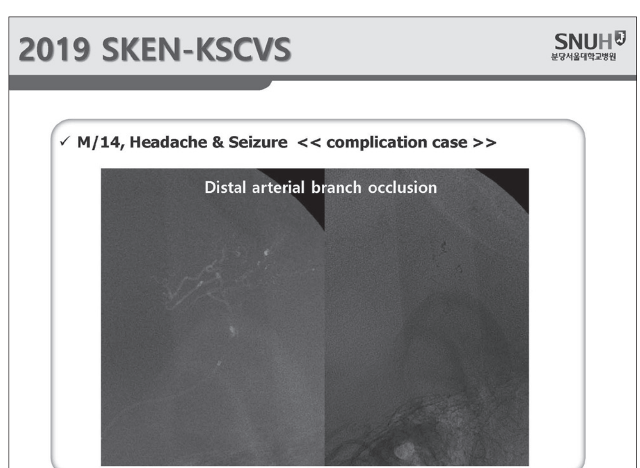

2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ F/54, headache
• 5yr-FU : no rebleeding
- No deficit


2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ F/30, coma


2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ F/30, coma
Thalamoperforating artery

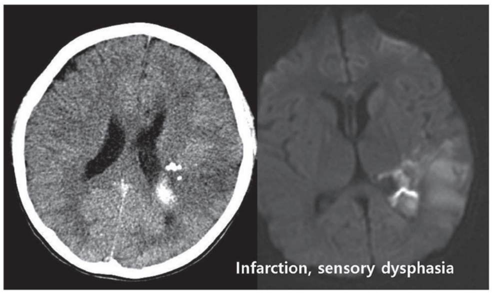

2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ F/30, coma
Post-embo
• 5yr-FU : no rebleeding
- Remained severe deficit


2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ M/14, Headache & Seizure << complication case >>


2019 SKEN-KSCVS
SNUH 분당서울대학교병원
✓ M/14, Headache & Seizure << complication case >>
Distal arterial branch occlusion

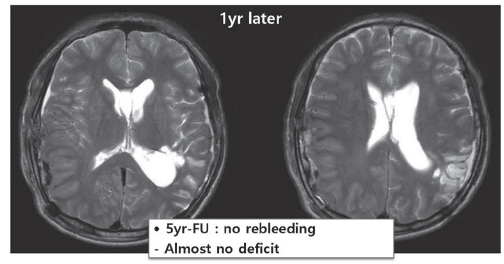
2019 SKEN-KSCVS

✓ M/14, Headache & Seizure << complication case >>



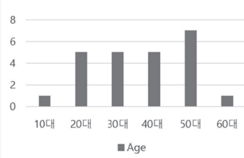
2019 SKEN-KSCVS

✓ M/14, Headache & Seizure << complication case >>



2019 SKEN-KSCVS

✓ 24 patients for 16 years



- Age : 41 ± 14 (14 ~ 62 yrs)
- Famale : 16 (64%)

- Prev. ischemic sx : 4
  - prev bypass op in 3 (1, 2, 6yrs)
- Perfusion study : perfusion decrease in 14, normal 4

- Prev. bleeding: 7
  - at the same site 4, 2 within 1 month
- Symptoms
  - headache, 15 / altered mentality, 9

2019 SKEN-KSCVS

✓ 24 patients for 16 years

- Location
  - LPChA, 11 / AChA, 9 / LSA, 3 / TPA, 1
- Laterality
  - Rt, 10 / Lt, 14
- Aneurysm size
  - 3.4 ± 2.1 mm (1.2 ~ 10.0 mm)
  - smaller than 2.5 mm in 50%
- Embolization materials
  - Glue (NBCA), 22 / Onyx, 2
- Occlusion technique
  - proximal occlusion, 9 / sac occlusion 15
  - all angiographically disappeared
- Date of embolization after bleeding
  - 23 ± 39 days (0 ~ 147), 54% within 1 week

2019 SKEN-KSCVS

✓ Results

- Complication
  - 2, new infarction from the occlusion (recovered)
- At follow-up
  - rebleeding at the same site : none
  - for 47 months (47 ± 35 months, 1 ~ 159 months)
  - hemorrhage in contralateral side in 3
- Recurred aneurysm
  - none, followed by TFCA (14), MRA (5), CTA (2), no FU study in 3
- Clinical outcome
  - normal life in 18 (75%)
  - moderate deficit in 3
  - bed ridden in 3 (as initial stage)

2019 SKEN-KSCVS

✓ Issue #1. Detection of the aneurysm

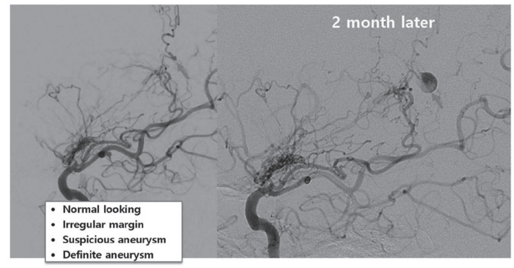
- Not all hemorrhage from MMD has an aneurysmal lesion. (1/3)
- But careful inspection is necessary
- More lesions can be found. (1/2)

2019 SKEN-KSCVS

✓ **M/28, Headache**

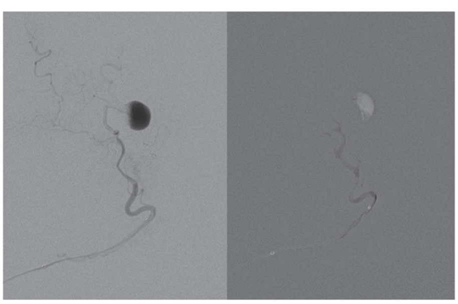
2019 SKEN-KSCVS

✓ **M/28, Headache**



2019 SKEN-KSCVS

✓ **M/28, Headache**



2019 SKEN-KSCVS

✓ **Issue #2. Indication**

- Not enough information on natural history
  - spontaneous regression
  - aggressive stance ?
  - no conclusion

2019 SKEN-KSCVS

✓ **Suggestion : when an aneurysmal lesion is detected**

- Endovascular occlusion is a feasible option
- Effective in prevention of rebleeding
- Not that difficult as it shows
- Not that risky as it shows

- Proximal occlusion seems to be enough
  - easier & safer option

2019 SKEN-KSCVS

Thank you very much.
감사합니다.

Special Lecture

좌장: 고준석 (경희대), 김종수 (성균관대)

1. Clinical application of DTI for stroke in the viewpoint of neurosurgery

장성호 (영남대 재활의학과)

Clinical application of DTI for stroke in the viewpoint of neurosurgery

장성호

영남대 재활의학과

Clinical Application of DTI for Stroke in Viewpoint of Neurosurgery

영남의대 재활의학교실

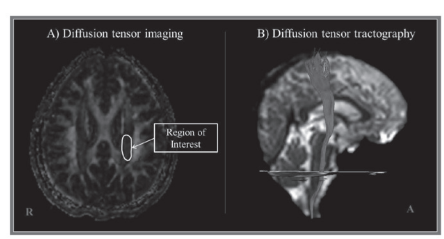
장 성 호

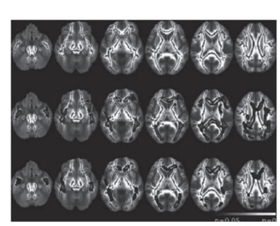
순서

- Diffusion tensor tractography
- Intracerebral hemorrhage
- Cranioplasty
- EVD
- Shunt

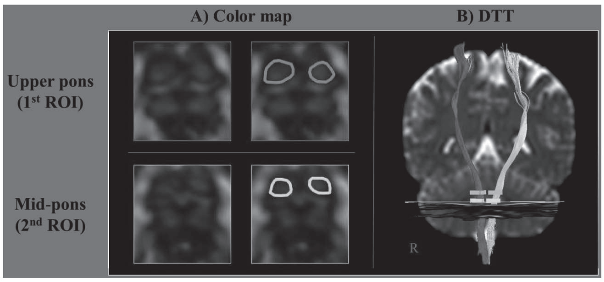
Our Brain Mapping Techniques

- Transcranial Magnetic Stimulation(TMS)
- Functional MRI
- **Diffusion tensor tractography (DTT)**
- Functional NIRS





Tract-based spatial statistics



Analysis conditions: 1) Fractional anisotropy, 2) Angle

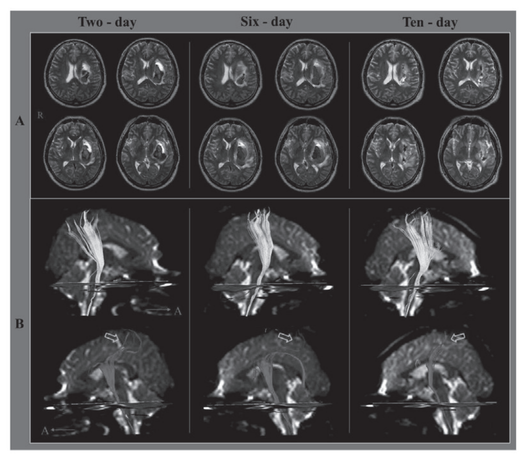
Repeatability & Reproducibility: 0.9~1.0

DTT로 reliable하게 분석할 수 있는 Neural tracts

- Motor: Corticospinal tract, Corticoreticulospinal tract, Corticofugal tract from secondary motor area, Corticobulbar tract
- Somatosensory: Medial lemniscus, Spinothalamic tract
- Movement: Dentatorubrothalmic tract, Corticopontocerebellar tract
- Consciousness: Ascending reticular activating system
- Cognition: Papez circuit, Fornix, Cingulum, Prefronto-thalamic tract, Prefronto-caudate tract
- Vestibular: Cortico-thalamo-vestibular tract, Vestibulo-cerebellar tract, Vestibulospinal tract
- Visual: Optic radiation
- Auditory: Auditory radiation, Auditory tract
- Language: Arcuate fasciculus
- Cranial nerves: CN 3, 5, 9, 10
- Others: Superior longitudinal tract, Inferior longitudinal tract, IFOF, Uncinate fasciculus, SCP, MCP, ICP, Corpus callosum

순서

- **Diffusion tensor tractography**
- **Intracerebral hemorrhage**
- **Cranioplasty**
- **EVD**
- **Shunt**



Difference of injury of the corticospinal tract according to surgical or conservative treatment in patients with putaminal hemorrhage

Table 2. Change of volumes of hematoma, peri-hematomal edema and total lesion.

	Surgical treatment group	Conservative treatment group
Hematoma volume on initial brain CT	30.69±4.16	29.17±3.43
Hematoma volume on brain MRI	15.41 ± 9.64^{a}	24.91 ± 9.45^{c}
Peri-hematomal edema volume on brain MRI	10.18 ± 9.81^{a}	26.80 ± 15.28^{c}
Total lesion volume on brain MRI	25.60±16.73	51.71 ± 20.05^{abc}

Average 23 days after onset

Difference of injury of the corticospinal tract according to surgical or conservative treatment in patients with putaminal hemorrhage

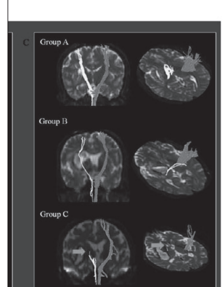
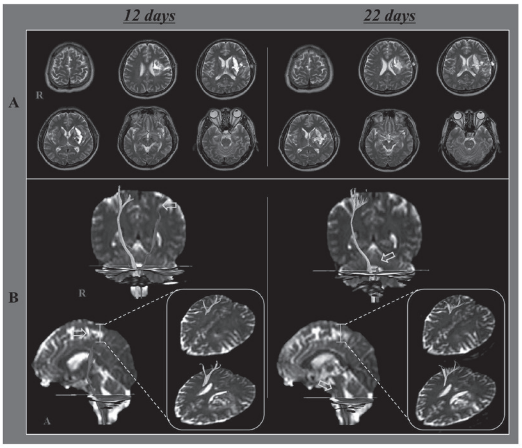


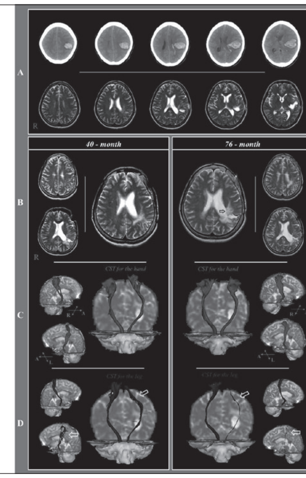
Table 3. Types of the affected corticospinal tract on diffusion tensor tractography.

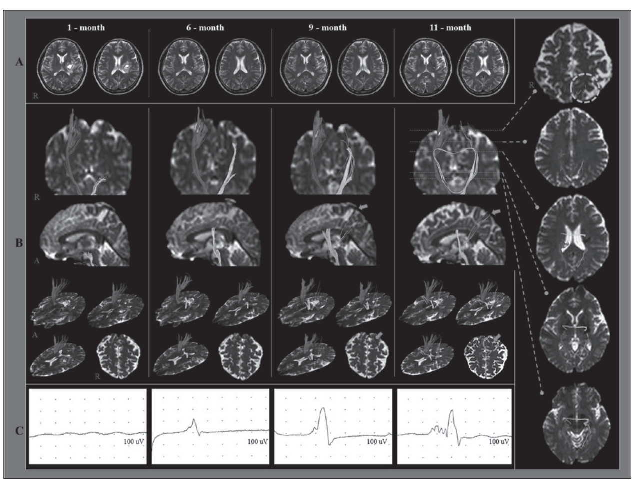
	Surgical treatment group ($n = 20$)	Conservative treatment group ($n = 26$)
Type		
Type A	8 (40%)	2 (8%)
Type B	2 (10%)	8 (31%)
Type C	10 (50%)	16 (61%)

Values are presented as the number of patients.



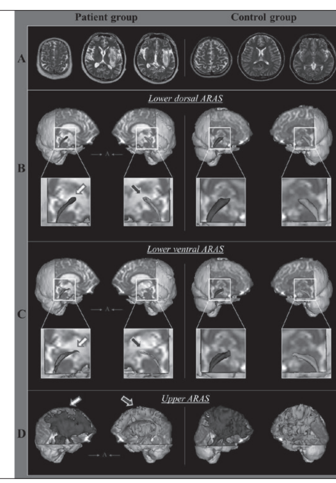
Delayed
Neuronal
Degeneration

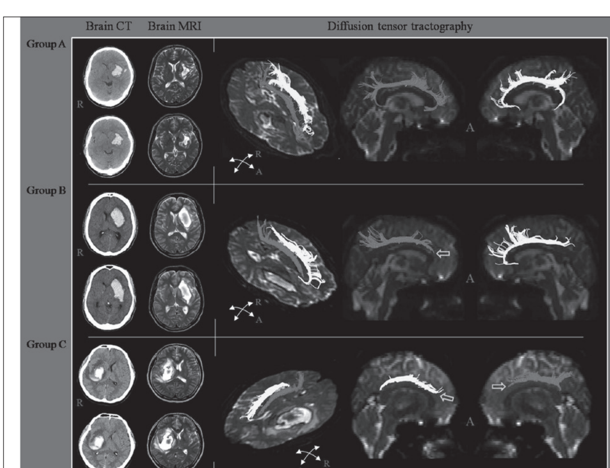




Impaired consciousness

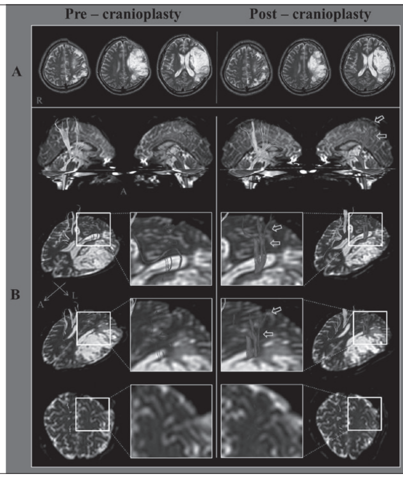
& affected lower dorsal ARAS

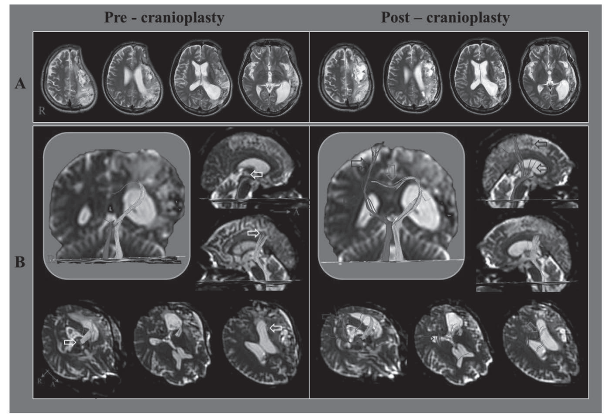


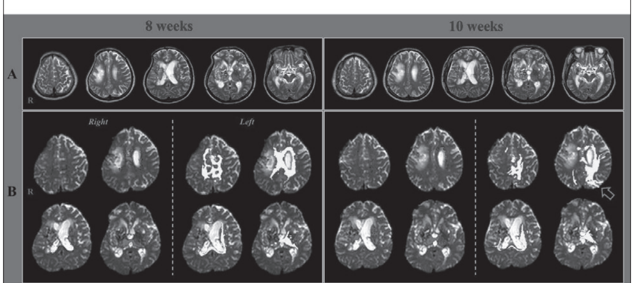


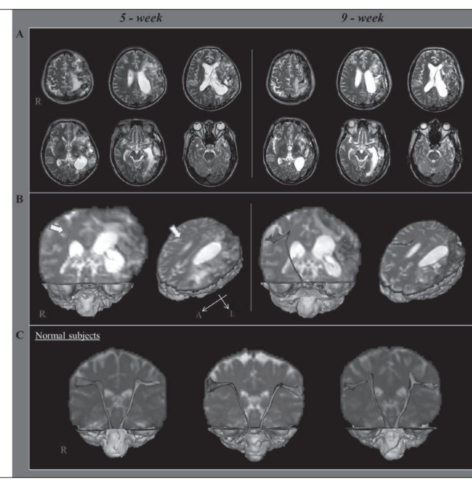
순서

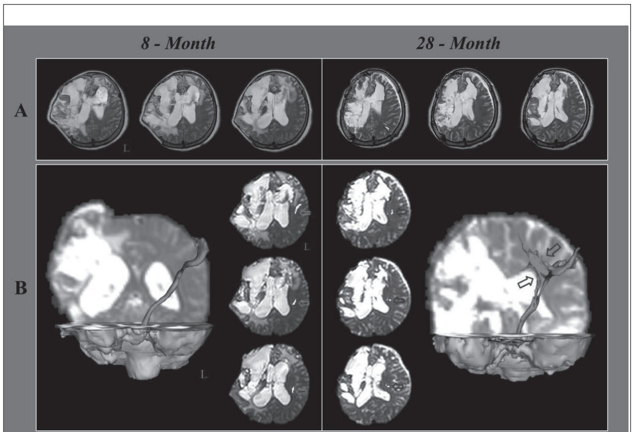
- **Diffusion tensor tractography**
- **Intracerebral hemorrhage**
- **Cranioplasty**
- **EVD**
- **Shunt**





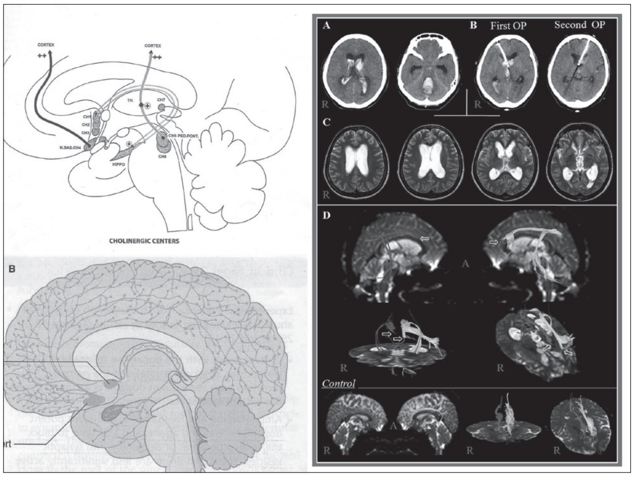


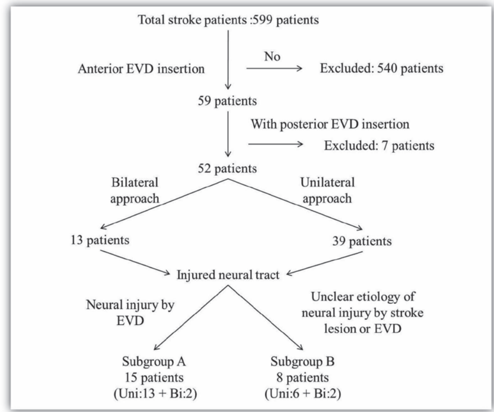




순서

- Diffusion tensor tractography
- Intracerebral hemorrhage
- Cranioplasty
- EVD
- Shunt





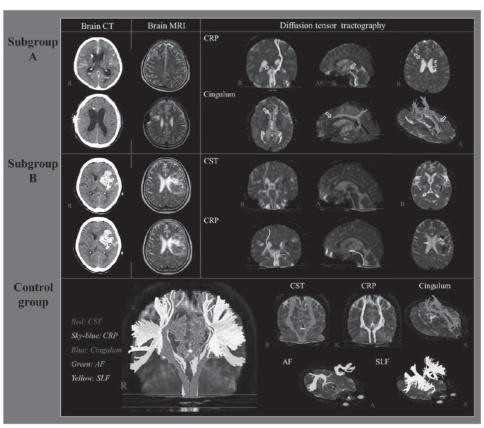
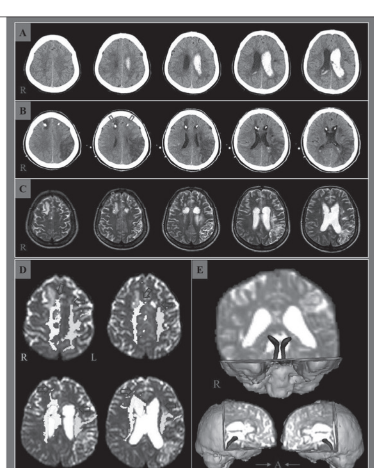
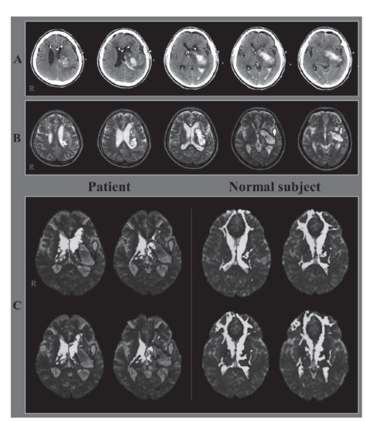


Table 1. Demographic and diffusion tensor imaging data of the patient and control group.

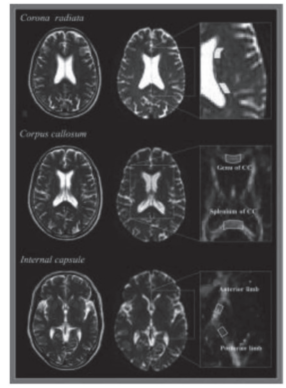
	Patient group ($n = 43$)	Injured group ($n = 15$)	Control group ($n = 22$)
Age	56.1 ± 9.7	58.4 ± 7.2	50.1 ± 16.1
Sex (Male/Female)	28/24	6/9	14/8
EVD approach (Unilateral/Bilateral)	40/12	13/2	
Diagnosis			
ICH+IVH+Hydro	5	0	
SAH+IVH+Hydro	16	8	
SAH+IVH	2	0	
ICH+IVH	13	5	
ICH+Hydro	4	0	
Other	3	2	
Incidence of neural injury			
CST	24 (55.8%)	0 (0.0%)	
CRP	14(32.6%)	7 (16.3%)	
Cingulum	12 (27.9%)	8 (18.6%)	
AF	2(4.7%)	0 (0.0%)	
SLF	2(4.7%)	0 (0.0%)	
Duration to DTI after EVD (days)	34 ± 12.8	30 ± 15.9	
Number of EVD;(one:two:three)	25:12:6	10:4:1	





순서

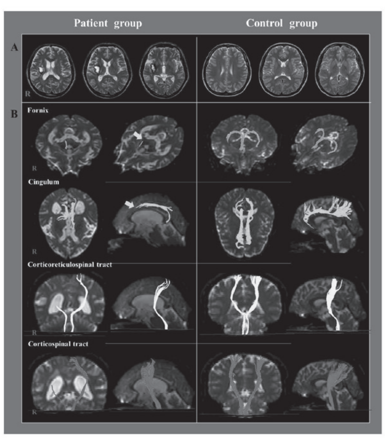
- Diffusion tensor tractography
- Intracerebral hemorrhage
- Cranioplasty
- EVD
- Shunt

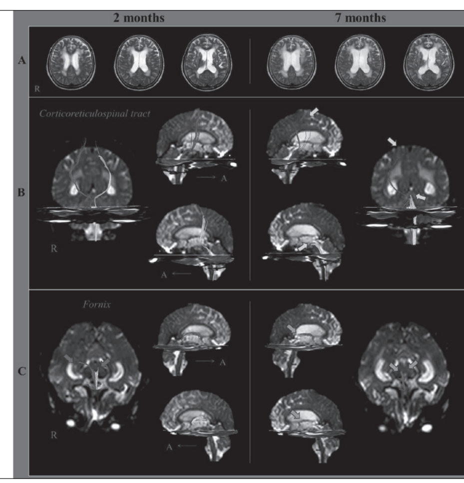


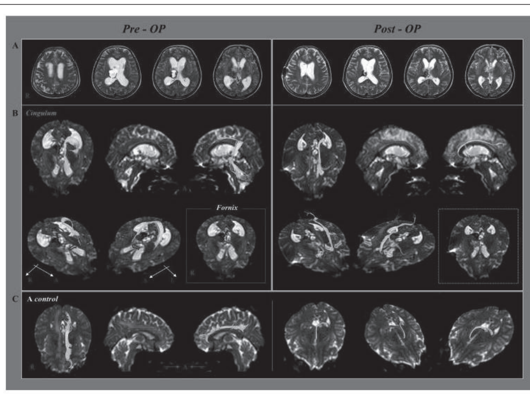
ICH

FA	Anterior CR	0.311 ± 0.117
	Posterior CR	0.504 ± 0.119
	Genu of CC	0.560 ± 0.122
	Splenium of CC	0.599 ± 0.117
	Anterior limb	0.413 ± 0.087
	Posterior limb	0.569 ± 0.139

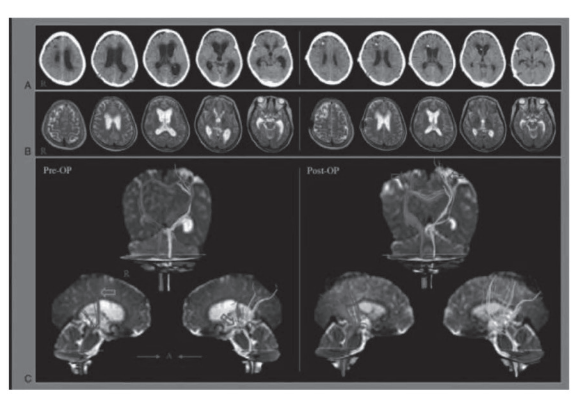
ICH



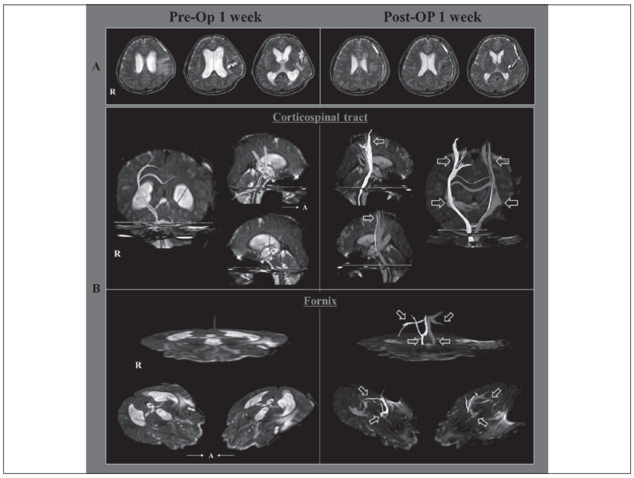


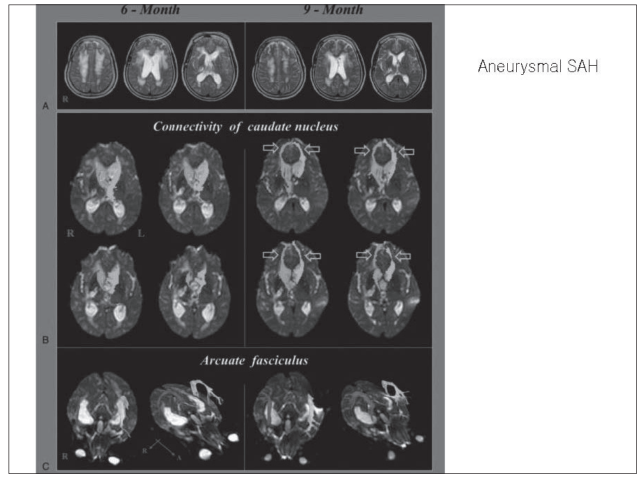


ICH, IVH, MMSE: 4-> 24(7days)



IVH Post-op 4 wk-walking


Pre-Op 1 week
Post-OP 1 week
A
R
Corticospinal tract
B
Fornix


6 - Month
9 - Month
Connectivity of caudate nucleus
Arcuate fasciculus
Aneurysmal SAH


감사합니다 !

Session III. Intracranial Dissection: General

좌장: 윤석만 (순천향대), 김태선 (전남대)

1. Epidemiology and pathophysiology of intracranial dissection
권오기 (서울대)

2. Diagnostic image of intracranial dissection
류창우 (경희대 영상의학과)

3. Natural course and medical treatment for intracranial dissection
김대원 (원광대)

Epidemiology and pathophysiology of intracranial dissection

권오기

서울대

Diagnostic image of intracranial dissection

류창우

경희대 영상의학과

두개강내 혈관박리는 비교적 젊은 연령에서 뇌졸중의 중요원인으로 인식되고 있다. 혈관 박리는 다양한 형태로 나타나며 일반적으로는 혈관내막(intima)이 손상되고 분리되면서 혈류에 밀려 혈관 중막(media) 및 외막(adventitia)으로 박리가 되면 협착, 폐색, 뇌동맥류 또는 뇌출혈으로 나타날 수 있다. 혈관박리에서 영상은 사실상 유일한 진단방법이며, intramural hematoma, double lumen, 박리성 동맥류, dissecting intimal flap 등을 영상에서 관찰하여 혈관박리로 진단하게 된다. 일반적으로Digital subtraction angiography가 표준 진단도구로 여겨지고 있으나, 혈관강의 외측을 못보는 한계가 있어 여러 다른 비침습적 영상방법을 병행하여야 진단이 가능한 경우가 있다.

이번 강좌에서는 두개강내 혈관박리의 영상진단의 기법과, 방법, 영상소견에 대하여 알아보고자 한다.

Digital Subtraction Cerebral Angiography

Digital subtraction cerebral angiography은 영상장비 중 동맥의 형태를 가장 정확히 보여줄 수 있고, 동적인 혈류를 보여주기 때문에, 아직까지 혈관박리의 진단에 있어서 골드스탠다드의 역할을 한다. 혈관조영술에서 두개강내 혈관박리의 소견은 1) intimal flap 2) double lumen 3) pearl and string sign 4) fusiform dilation and stenosis등이다. Intimal flap과 double lumen은 특징적인 소견이나 일부의 경우에서만 관찰되며 Pseudolumen은 명확한 경계가 없이 조영제의 정체만으로 보일 수 있다. pearl and string sign과 fusiform dilataion and stenosis는 비교적 흔하게 볼 수 있는 소견이나 이러한 두 소견이 혈관박리 외의 다른 경우에도 볼 수 있는 소견이기 때문에 확진의 소견으로 간주할 수 없다. 이러한 이유로 근래에 들어서는 MRI 및 MRA가 혈관박리가 의심되는 환자에서 중요한 일차영상검사로 여겨지고 있다.
이와 같이 혈관조영술로 진단이 명확치 않은 경우 다른 영상방법을 병행하여 혈관강 바깥의 소견, 즉 intramural hematoma와 pseudolumen을 관찰하여야 정확한 진단을 할 수 있다.

CT Angiography

CT angiography (CTA)는 빠르게 뇌혈관의 영상을 얻을 수 있는 장점이 있는 검사이며, 기본적으로 두개강내 혈관박리의 CTA에서는 고식적 혈관조영술과 같이 intimal flap 과 double lumen을 봄으로써 혈관박리를 진단하게 된다. CTA의 source images에서 혈관강 외측의 병리를 단면으로 볼 수 있기 때문에 pseudolumen을 관찰할 수 있다. 그러나 wall thickening으로만 보이는 소견은 동맥경화증과 감별하기 어려울 수 있다. 또한 영상획득에 있어 적절한 타이밍의 스캔과 조영제의 투여속도가 요구되기 때문에 진단에 적합한 영상을 얻지 못할 수도 있다.

MRI and MRA

Intramural hematoma는 특징적으로T1 강조영상에서 고신호강도의, 초생달 모양의 혈관벽의 두꺼워짐으로 관찰된다. 또는 혈관벽의 두께 증가는 positive remodeling의 양상으로 나타난다. 자주 보이는 소견은 아니지만 단면영상 및 MRA source imaging에서 intimal flap이 관찰될 수도 있다. DSA와 같이 MRA에서도 pearl and string sign과 fusiform dilation and stenosis 로 관찰될 수 있다. 그러나 MRI, MRA는 해상도가 conventional angiography보다 낮기 때문에 단순한 atherosclerosis의 소견으로 오해하고 진단이 안될 수도 있다. 또한 time-of-flight (TOF) MRA검사의 특성상 혈관내 turbulence flow에 의해 intimal flap이 위양성으로 인식될 수 있다. Peri-arterial fat이나 intraluminal thrombus 역시 Intramural hematoma로 오인될 수 있다. 그 외 fenestration과 같은 해부학적 변이도 혈관박리로 오진할 수 있다. 이러한 단점을 보완하기 위하여 다음과 같은 추가적인 영상검사방법이 추천되고 있다.

1. 조영증강 MRI 및 MRA검사: intimal flap과 double lumen을 더 용이하게 진단할 수 있도록 해준다.

2. Susceptibility weighted imaging: 혈액성분을 강조하여 보여주는 영상이며 intramural hematoma가 crescent dark rim으로 보인다.

3. Basiparallel Anatomic Scanning (BPAS): vertebrobasilar artery의 장축에 따라 20mm의 절편으로 두꺼운 영상을 획득하여 혈관외벽을 형상화 하는 기법으로 혈관외벽과 MRA의 혈관내 형태를 비교함으로써 혈관박리를 진단할 수 있다.

Recommendation of Diagnostic Procedures for Intracranial Dissection

출혈성 박리

급성 지주막하 출혈이 동반된 혈관박리는 CTA에서 double lumen, pearl and string sign, fusiform aneurysm을 진단하고, saccular aneurysm을 배제하는 것이 필요하다. 이후 DSA를 시행하여 이러한 소견을 확진하고 혈관내 치료의 가능성을 확인하여야 한다.

허혈성 박리

MRI가 아주 중요한 진단 수단이며 posterior circulation의 경우 BPAS와 MRA를 동시에 얻어 비교하는 방법이 효과적이다. MRA에서 double lumen, intramural hematoma, fusiform dilatation, and the pearl and string sign을 진단하고 T1강조 영상과 MRA source 영상에서 혈관벽의 intramural hemorrhage 와 intimal flap을 진단할 수 있다. 조영증강 T1 강조영상 또는 MRA source 영상에서는 double lumen을 더 쉽게 진단할 수 있다.

영상검사의 한계

혈관박리의 진단 자체가 영상검사에 전적으로 의지하는 질환이기 때문에 영상검사에 대한 의존도가 높다. 그러나 고령에서 전형적인 증상이 없이 발생한 혈관박리의 경우, 영상 검사의 소견이 atherosclerosis와 감별이 어려울 수 있다. 이러할 경우 혈관박리는 시간에 따라 변하기 때문에 추적검사가 중요하다.

결론

혈관박리는 급성기에 적절히 진단이 되지 않고 치료가 늦어진다면 환자에게 심각한 결과를 초래할 수 있다. 따라서 빠르고 정확한 진단이 필요하며 이를 위해 다양한 방법의 영상검사법과 영상소견을 이해하는 것이 필요하다.

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Natural course and medical treatment for intracranial dissection

김 대 원

원광대

Although intracranial artery dissection is less common than cervical artery dissection in adults of European ethnic origin, intracranial artery dissection is reportedly more common in children and in Asian populations. Noninvasive neuroimaging techniques are increasingly identifying unruptured intracranial arterial dissections (IADs) at examination for headache or ischemic symptoms. Approximately 3% of cases of aneurysmal subarachnoid hemorrhage (SAH) are caused by IADs, but the natural history of unruptured IADs is not known. Intracranial artery dissections (IADs) are still considered relatively rare but are an important cause of stroke, especially in young adults. Furthermore, they are a cause of subarachnoid hemorrhage (SAH) or compression on brainstem or cranial nerves and are therefore of clinical significance. Publications on IAD presenting with ischaemia are scarce and comprise small series, whilst there are many publications about IAD presenting with SAH including large series. As a consequence, these studies are prone to recruitment bias. General recommendations are therefore difficult to formulate and it seems necessary to distinguish between clinical presentations with SAH or ischaemia since the natural history and prognosis of these conditions are very different. Treatment of intracranial artery dissections is empirical in the absence of data from randomised controlled trials. Most patients with subarachnoid hemorrhage undergo surgical or endovascular treatment to prevent rebleeding, whereas patients with intracranial artery dissection and cerebral ischaemia are treated with antithrombotics. Prognosis seems worse in patients with subarachnoid hemorrhage than in those without. Currently, there are no studies supporting the superiority of anticoagulants to antiplatelet agents. A recent study failed to show a risk reduction of recurrent ischaemic stroke in patients receiving anticoagulants compared with those treated with antiplatelet agents. The use of anticoagulants in the acute phase of IAD is controversial as it can lead to progression of the dissection, expansion of the intramural hematoma and increased luminal stenosis or hemorrhagic transformation of recent cerebral ischemia. However, treatment with antiplatelet agents or anticoagulants appears to be safe according to some cohort studies. It is recommended to maintain anticoagulants or antiplatelet agents for at least 3–6 months or until complete recanalization of the artery has occurred. Patients with recurrent or progressive ischemic symptoms, despite medical therapy, or with enlargement of the dissecting aneurysm are usually treated with endovascular or surgical approaches.

Session IV. Intracranial Dissection: Treatment

좌장: 신용삼 (가톨릭대), 정영균 (인제대)

1. Surgical treatment for intracranial dissection
박재찬 (경북대)

2. Endovascular treatment for intracranial dissection
백승국 (부산대 영상의학과)

3. Complicated cases and panel discussion
주성필 (전남대)
김성림 (가톨릭대)
이재일 (부산대)
박중철 (울산대)

Surgical treatment for intracranial dissection

박재찬

경북대

This presentation is particularly about the surgical treatment of dissecting anterior circulation intracranial aneurysms. Cerebral aneurysms arising at the non-branching sites are classified into 4 different types based on the lesional patterns of the internal elastic lamina. As Mizutani type 1, classic dissecting aneurysms of intracranial carotid circulation are extremely rare, they are discussed based on the clinical study by Ohkuma et al. Characteristic angiographic findings to confirm Mizutani type 1 arterial dissection are as follow: (1) the double-lumen sign (the presence of false lumen or an intimal flap) is used as a reliable finding of arterial dissection; (2) stenosis with dilatation (the pearl and string sign) is also used as a reliable finding of arterial dissection; (3) dilatation without stenosis requires an additional definition to confirm arterial dissection (Discoloration of the affected artery around the aneurysmal dilatation, which is considered to be due to intramural hematoma, should be seen during operation); and (4) stenosis without dilatation (the string sign) or occlusion (tapered occlusion) also requires additional definitions. Stenosis is not segmental but extensive on initial angiography, and resolution of stenosis or occlusion should be seen on follow-up angiography, which is considered to be a reliable angiographic sign of arterial dissection. The patient groups of bleeding presentation and ischemic presentation are compared in their angiographic findings and treatment.

Mizutani type 4 aneurysms arise in arterial areas with minimally disrupted internal elastic lamina without intimal thickening. In the presenter's clinical series, blister-like aneurysms arising at the internal carotid artery (n = 20), anterior communicating artery (n = 4), and middle cerebral artery (n = 3) are discussed. All blister-like aneurysms presented with acute subarachnoid hemorrhage and were treated surgically. Meanwhile, a case of an intra-arterial schwannoma at the M1 segment of a middle cerebral artery is discussed. It was misdiagnosed as an arterial dissection preoperatively, presenting with a subarachnoid hemorrhage and a small bulge of the M1 segment on the preoperative angiogram.

Endovascular treatment for intracranial dissection

백승국

부산대 영상의학과

I. INDICATION

First of all, patients presenting with an SAH should be considered endovascular treatment, particularly those with intracranial artery dissections (IAD) in the posterior circulation. These patients should first undergo a diagnostic angiogram to assess for a direct cause of the hemorrhage, such as a pseudoaneurysm. The diagnostic angiogram would also study the collateral circulation, the length of the dissected segment, and vertebral dominance. Intracranial pseudoaneurysms have a reported mortality rate as high as 83% and a rerupture rate of approximately 40 - 70% within the first 24 hours of presentation. Occlusion of the entire dissected segment with or without flow replacement is the treatment option associated with the lowest risk of rehemorrhage. Although this could be achieved through proximal occlusion, trapping of the dissected segment might be a better option to avoid filling of the occluded segment through a retrograde flow from the contralateral vertebral artery. The procedure can be performed endovascularly, although in certain cases an open surgical approach might be preferable to visualize and preserve the PICA and brainstem perforators.In cases of ruptured vertebral artery dissection that extends to the basilar artery, treatment options are limited. These lesions are notoriously difficult to treat. If the patient passes the balloon test, occlusion and then a proximal occlusion of the basilar artery or bilateral occlusion of the vertebral arteries might be the best option. Otherwise, a stent-in-stent technique can be attempted with potential stent coiling of the pseudoaneurysm.

Another indication in which endovascular treatment is indicated in the acute setting is when dissection results in acute arterial occlusion. In these cases, hemodynamic compromise, and not thromboembolism, is the mechanism behind ischemia of cerebral tissue. In these patients, attempts can be consistent to restore flow. First, high-volume fluid resuscitation should be started to reach a normotensive state with a likely benefit of a slight hypertensive state. Once the patient is stabilized, an intervention to restore normal flow should be considered.

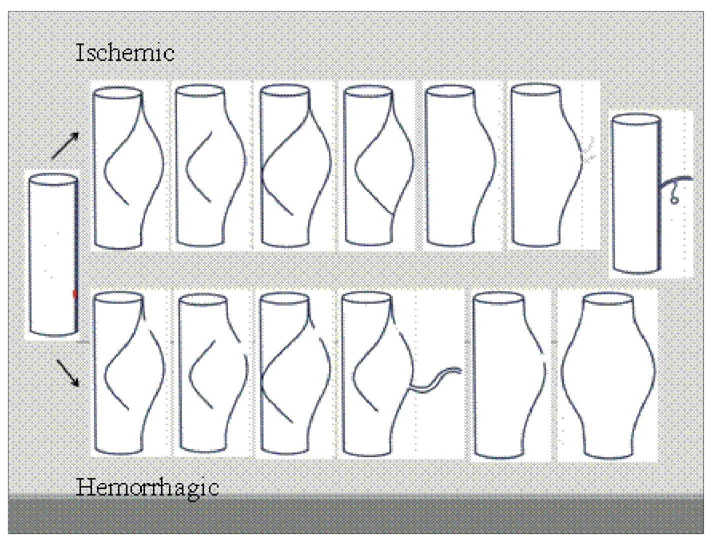
Other than these 2 indications, endovascular therapy should be considered if medical management fails or the patient has a contraindication to antithrombotic agents. Although no textbook definition of failure of medical therapy exists, it is generally agreed that progression of neurologic symptoms and new ischemic events despite adequate antithrombotic therapy define failure of medical therapy. Whether pseudoaneurysm enlargement can be considered failure of medical therapy is controversial.

II. MANAGEMENT

Ischemic

Endovascular treatment is undertaken only in patients with recurrent ischemic symptoms despite receiving optimum medical treatment. Sometimes, endovascular treatment is undertaken if the dissecting aneurysm has increased in size, to prevent rupture, or more rarely to reduce signs of brainstem compression. In children, the preferred and widespread practice is surgical or endovascular treatment in patients with intracranial artery dissection with subarachnoid hemorrhage and those without subarachnoid hemorrhage and mass effect, whereas patients with intracranial artery dissection without subarachnoid hemorrhage and cerebral ischemia tend to be given medical treatment

Treatment options include intra-arterial thrombolysis, coils, and stents. Endovascular therapies are not only minimally invasive but they offer the ability to both diagnose and treat patients within a single procedure and are thus being adopted into practice more broadly. By allowing for the direct delivery of thrombolytic agents at the site of dissection, intra-arterial thrombolysis is preferred to intravenous thrombolysis because it decreases the resulting dose requirements and reduces the risk of complications. Intra-arterial thrombolysis in conjunction with either a bypass or a stent has also been shown to be effective. Endovascular therapies include the deconstructive and reconstructive treatments. The details will be described below.

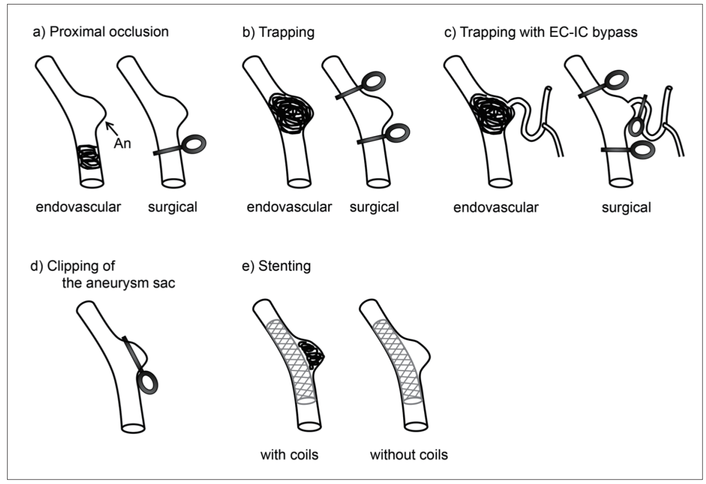


Hemorrhagic

Because of the risk of rebleeding and higher mortality, IAD in patients presenting with hemorrhage are treated more aggressively than those involving ischemia. Patients who are treated conservatively for SAH have been found to have poor clinical outcomes, resulting from frequent rebleeds. Ruptured dissections are unstable and have a tendency to rebleed. Studies on the treatment of posterior circulation IADs presenting with SAH report high mortality and poor outcomes. However, this situation is often confounded by the poor initial presentation and preexisting contraindications to more aggressive therapies.

Endovascular therapies include the deconstructive and reconstructive techniques including proximal occlusion, trapping with or without extracranial-intracranial (EC-IC) bypass, coiling of the aneurysm sac, stent-assisted coil embolization of the aneurysm including the use of flow diverters.

These options are individually chosen for patients based on vessel morphology and anatomy (i.e., the involvement of critical artery or perforating branches).



1. Parent artery occlusion (Fig. a, b, c) is a deconstructive technique in which blood flow into the dissected segment of the artery is stopped by occlusion either surgically or through an endovascular approach. Preferably, the dissected segment is occluded both proximally and distally to prevent rerupture through retrograde filling of a dissecting aneurysm. Parent artery occlusion has a risk of brain infarct in case of insufficient collateral supply. Before permanent occlusion, the collateral supply can be assessed with various methods.

2. Reconstruction (Fig. d, e) can be performed using flow diversion devices or stents to separate the true and false lumens. This strategy leads to a controlled thrombosis of the dissection or the dissecting aneurysm and preserves patency of the parent vessel, thereby preserving physiologic cerebral blood flow. In endovascular treatment, stent-assisted coiling can be used. Stenting of the dissected artery without any coiling by flow-diverter stents or conventional close-cell stents has been reported in small series; however, several days to months can pass before the dissecting aneurysm is thrombosed. Moreover, stenting needs dual antiplatelet treatment for several months after the procedure, thereby exposing patients to an increased risk of hemorrhagic complications.

3. Endovascular treatment of IAD is typically performed with coils and/or stent including flow diverters. However, in the certain cases such as peripheral located dissecting aneurysm or severe luminal narrowing within the diseased segment may preclude distal access for coiling or stenting. In this situation, a liquid embolic agent such as glue or Onyx may be helpful for traversing the stenosis and achieving parent artery occlusion or aneurysm obliteration.

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Complicated case 1

주성필

전남대

Complicated case

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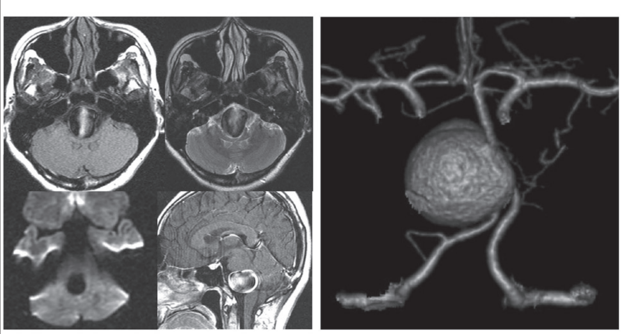
Case illustration

- A 27 year old girl
- C/C : dizziness
- N/E : Alert m/s
  Pupil 3+/3+
  No motor weakness

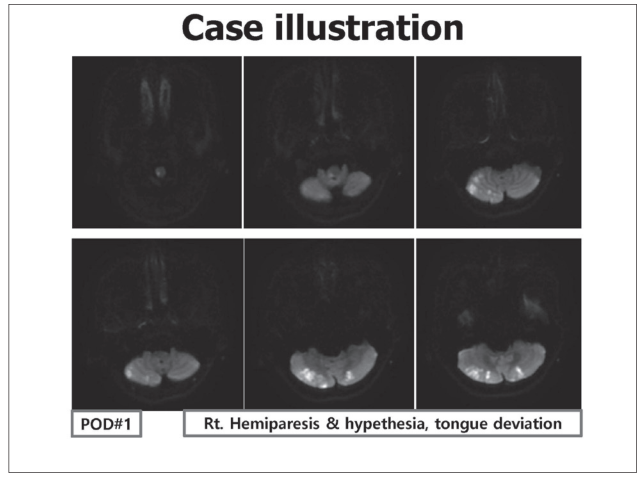
  Higher cortical function : W.N.L.
  C.N. exam : W.N.L.
  Cbll sign : W.N.L.

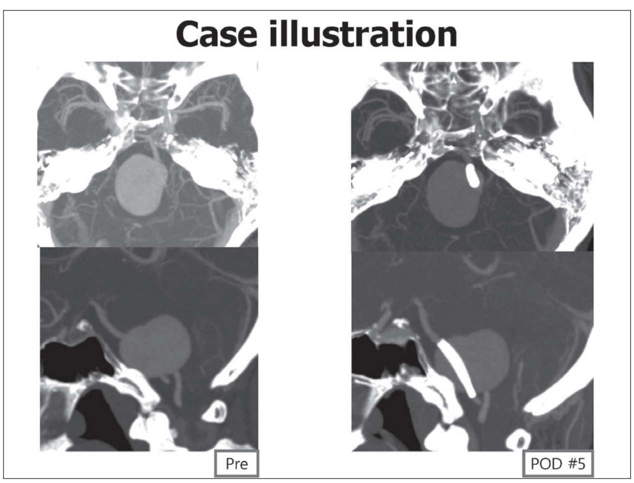
Case illustration

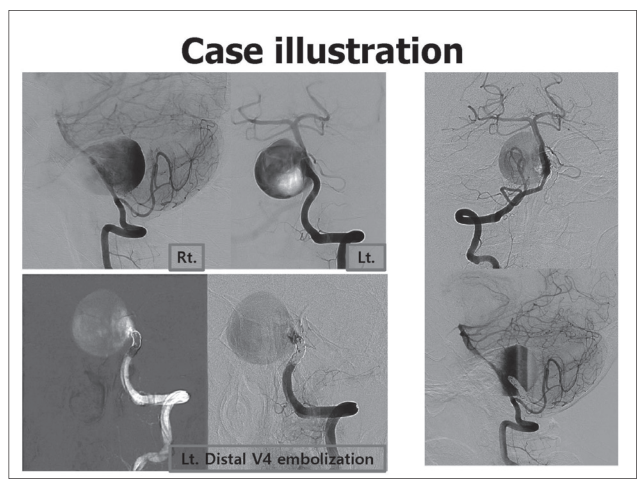


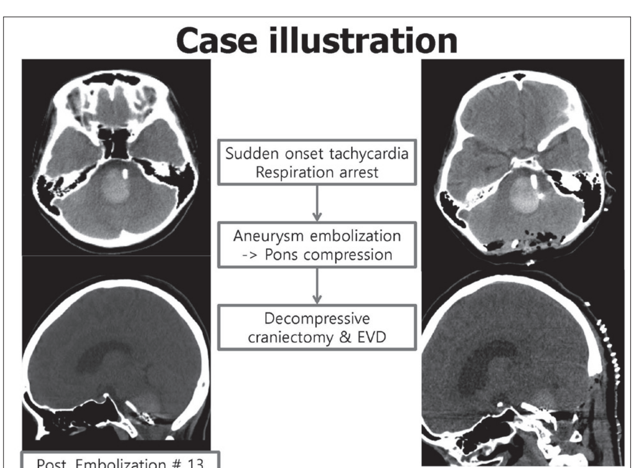
Case illustration

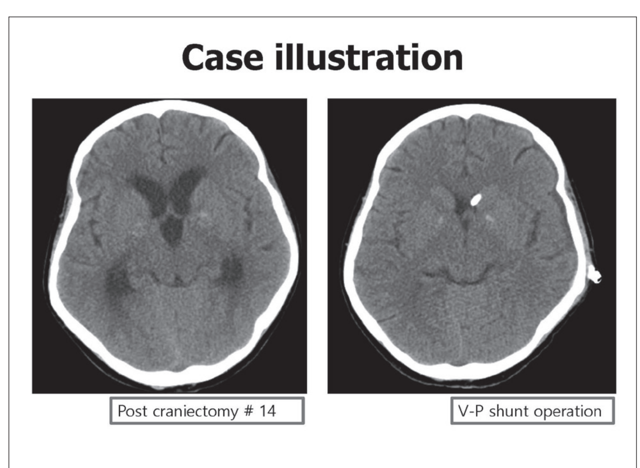

Case illustration


Case illustration
POD#1
Rt. Hemiparesis & hypethesia, tongue deviation

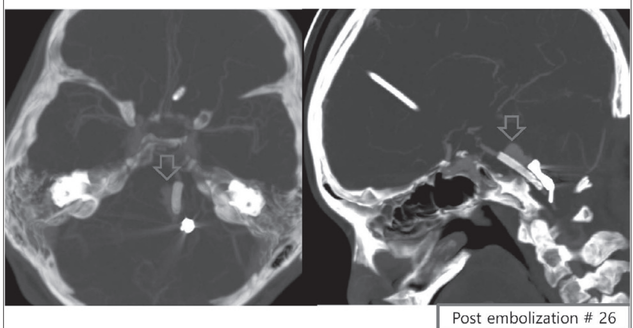

Case illustration
Pre
POD #5


Case illustration
Rt.
Lt.
Lt. Distal V4 embolization

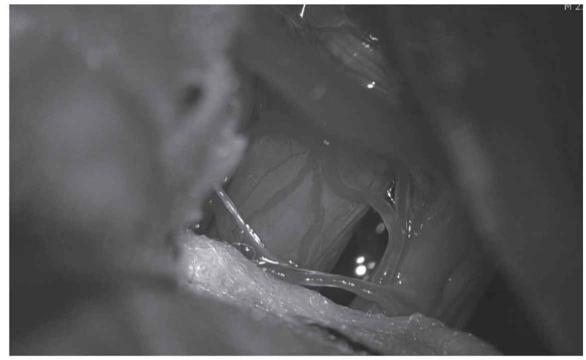

Case illustration
Sudden onset tachycardia
Respiration arrest
Aneurysm embolization
-> Pons compression
Decompressive
craniectomy & EVD
Post. Embolization # 13


Case illustration
Post craniectomy # 14
V-P shunt operation

Case illustration



Vasa Vasorum: Another factor ?



Complicated case 2

김성림

가톨릭대

Complicated case 3

이 재 일

부산대

Fibromuscular dysplasia (FMD) is a noninflammatory arterial disease with female dominancy. The arterial manifestrations could include dilatation, stenosis, dissection, aneurysm or vessel tortuosity. We reported an extremely rare case of subarachnoid hemorrhage caused by internal carotid artery (ICA) dissecting rupture in a patient with fibromuscular dysplasia.

A 38-year-old female was admitted to our neurointensive care unit with a history of sudden onset severe headache. A CT demonstrated subarachnoid hemorrhage at right sylvian fissure and basal cistern. A CT angiography showed tiny dilatation of right ICA communicating segment and left proximal ICA dissection. Conventional angiography confirmed right distal ICA dissection and Lt proximal ICA dissection with distal beads on a string sign. One day follow-up angiography revealed vessel wall remodeling, but we performed multiple stenting at dissecting segment considering risk of rebleeding. Double stenting facilitated dissecting flap reposition and reduced tiny dilatation portion. Seven days follow up angiography showed no further aneurysmal dilatation or in-stent stenosis and 3 months follow DSA confirmed complete obliteration of aneurysmal dilatation without stenosis.

We should consider FMD in young female patient with hypertension history and intracranial aneurysms or dissection. We also bring up the diagnosis and discussion of a cervico-encephalic artery and renal artery examination during the diagnostic work-up.

Complicated case 4

박중철

울산대

MEMO

대한뇌혈관내수술학회 | 대한뇌혈관외과학회

SKEN-KSCVS 합동 연수강좌

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발 행	2019년 9월 7일
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