Twenty seven years Experience in the Treatment of Moyamoya Disease (1982-2007)

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Chile
Twenty seven years Experience in the Treatment of Moyamoya Disease (1982-2007)

- Chronic cerebrovascular occlusive disease
- Arteritis (endarteritis)
- Genetic immunological base
- Name: “cigarette smoke in the air”

Moyamoya in Japanese

Jiro Suzuki
PROGRESSIVE STENOSIS OF INTRACRANIAL INTERNAL CAROTID ARTERIES , ACA & MCA
PROGRESSIVE APPEARANCE OF A MESH OF VESSELS OF SMALL DIAMETER IN THE REGION OF BASAL GANGLIA, AS A FORM OF COLLATERAL CIRCULATION

Stages of progression
ISCHEMIC FORM

- Childs
- T.I.A
- Infarction
- Cerebral atrophy

HEMORRHAGIC FORM

- Young adult
- Ventricular hemorrhage
What is the meaning of moyamoya vessels?
Conclusions: These results demonstrated that chronic cerebral hypoperfusion could induce sustained up-regulation of VEGF mRNA and protein expression in rat brain, which was correlated with angiogenesis.

Enhanced brain angiogenesis in chronic cerebral hypoperfusion after administration of plasmid human vascular endothelial growth factor in combination with indirect vasoreconstructive surgery

Conclusions. In rat models of chronic cerebral hypoperfusion, administration of phVEGF combined with indirect vasoreconstructive surgery significantly increased capillary density in the brain. The authors’ results indicate that administration of phVEGF may be an effective therapy in patients with chronic cerebral hypoperfusion, such as those with moyamoya disease.
ISCHEMIA
HYPOXIA

HIF

VEGF

PDGF

VEGFR

PDGFR

cell elected

ANGIOGENESIS
Twenty seven years Experience in the Treatment of Moyamoya Disease (1982-2007)

15 cases (7 males, 8 females. Age: 6 – 46 y.o; media: 25,4 y. old)

Ischemic cases
7 cases; 6 typical, 1 atypical

<table>
<thead>
<tr>
<th>Name</th>
<th>Age</th>
<th>Gender</th>
<th>StrokeType</th>
<th>Moyamoya clinical pattern</th>
</tr>
</thead>
<tbody>
<tr>
<td>R.A.</td>
<td>6</td>
<td>M</td>
<td>Ischemic</td>
<td>Typical</td>
</tr>
<tr>
<td>L.M.</td>
<td>8</td>
<td>M</td>
<td>Ischemic</td>
<td>Typical</td>
</tr>
<tr>
<td>J.N.</td>
<td>9</td>
<td>F</td>
<td>Ischemic</td>
<td>Typical</td>
</tr>
<tr>
<td>Y.M.</td>
<td>11</td>
<td>M</td>
<td>Ischemic</td>
<td>Typical</td>
</tr>
<tr>
<td>L.O.</td>
<td>15</td>
<td>M</td>
<td>Ischemic</td>
<td>Atypical (unilat)</td>
</tr>
<tr>
<td>F.G.</td>
<td>16</td>
<td>M</td>
<td>Ischemic</td>
<td>Typical</td>
</tr>
<tr>
<td>F.C.</td>
<td>16</td>
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Twenty seven years Experience in the Treatment of Moyamoya Disease (1982-2007)

15 cases (7 males, 8 females. Age: 6 – 46 y.o; media: 25.4 y. old)

Hemorrhagic cases
8 cases; 3 typical, 5 atypical

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</tr>
<tr>
<td>L.E.</td>
<td>25</td>
<td>F</td>
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<td>Typical</td>
</tr>
<tr>
<td>H.N.</td>
<td>36</td>
<td>M</td>
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<td>Atypical (unilat)</td>
</tr>
<tr>
<td>V.S.</td>
<td>37</td>
<td>F</td>
<td>Hemorrhagic</td>
<td>Atypical (unilat)</td>
</tr>
<tr>
<td>R.C.</td>
<td>42</td>
<td>F</td>
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Angiographical criteria to diagnose the Moyamoya Disease
(Reduction committee on progressive occlusive disease of the circle of Willis, Ministry of Health and Welfare of Japan 1978)

1- Stenosis or occlusions at the terminal portion of intracranial ICA and/or the proximal portion of the ACA and/or the MCA.
2- Abnormal vascular mesh, the “moyamoya” vessels, observed in the neighborhood of the above mentioned areas in the arterial phase.
3- Above mentioned findings are recognized bilaterally.

We observed 6 atypical cases (CUASI MOYAMOYA) or Akim Moyamoya (40%)

4 cases Hemorrhagic Stroke – unilaterally moyamoya vessels
1 case Hemorrhagic Stroke – early occlusion of cervical ICA bilat.
1 case Ischemic Stroke – unilaterally moyamoya vessels
CASE Nº 6  J.G. 18 years old, female
Mild headache, left hemiparesis, with complete recovery.
CASE Nº 2  J.N. , 9 years old, female
Difficult learning, speech disturbance, right progressive hemiparesis, focal convulsions.
ENCEPHALO-DURO-ARTERIO-SINANGIOSIS (EDAS)
1. ENCEPHALO-DURO-ARTERIO-GALEO-SINANGIOSIS
Burr Holes – Periostial -Sinangiosis
ENCEPHALO-DURO-ARTERIO-GALEO-SINANGIOSIS
BURR HOLES – PERIOSTIAL -SINANGIOSIS
ANGIOGRAPHICAL CONTROL (6-12 months)
MEDICAL TREATMENT

Vasodilators drugs
Corticosteroids
Platelets antiaggregants
Nootropics drugs

NO UTILITY DEMONSTRATED

SURGICAL TREATMENT

10 first operations (1982-2000) : EDAS

5 last operations (2001-2007) : EDAS + wide galeal flap
Encephalo-duro-galeo-arterio-sinangiosis
+burr holes and periostial sinangiosis
<p>| | |</p>
<table>
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<tr>
<td>I</td>
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</tr>
<tr>
<td>II</td>
<td>Activity with mild limitations</td>
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<tr>
<td>III</td>
<td>Activity with severe limitations</td>
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<tr>
<td>IV</td>
<td>Vegetative state</td>
</tr>
<tr>
<td>V</td>
<td>Died</td>
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## ANALYSIS OF FUNCTIONAL RESULTS ACCORDING WITH THE INITIAL DAMAGE

<table>
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<tr>
<th>Ischemic cases</th>
<th>Hemorrhagic cases</th>
<th>Grade</th>
</tr>
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<tbody>
<tr>
<td>2 TIA</td>
<td>1 small ICH ; 1 IVH</td>
<td>I</td>
</tr>
<tr>
<td>3 Infarction (2 temp; 1 front.)</td>
<td>1 IVH; 1 IVH + lobar ICH</td>
<td>II</td>
</tr>
<tr>
<td>1 Hemispheric Infarction</td>
<td>2 severe IVH</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2 IVH + basal ganglia ICH</td>
<td>III</td>
</tr>
<tr>
<td>1 Bilateral multiple infarctions</td>
<td></td>
<td>IV</td>
</tr>
</tbody>
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**Initial clinical presentation and time of evolution of Moyamoya Disease is strongly related with the final outcome**
RESULTS AT 6 TO 12 MONTHS AFTER OPERATION

Clínical results

I  Return to normal life  4 cases  
II Activity with mild limitations  5 cases  
III Activity with severe limitations  5 cases  
IV Vegetative state  1 case  
V Death  0 cases  

Angiographical results

14/15 Presented signs of effective revascularization
1/15 Not presented signs of effective revascularization

The revascularization signs were present between 6-12 months after the operation, and the donnor artery with wider caliber
## OUTCOME
Follow up between 2 – 27 years

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Related with:

1- Early Diagnosis
2- Effective treatment

Outcome related with:
- Early Diagnosis
- Effective treatment

**Outcome Rating:***
- Excellent
- Good
- Poor
CONCLUSIONS

1- THE PATTERN OF MOYAMOYA DISEASE IS SOMEWHAT DIFFERENT IN OUR EXPERIENCE, FROM THE JAPANESE EXPERIENCE, WITH MANY CASES CATALOGUED AS “CUASI MOYA MOYA”, PERHAPS WE SHOULD SAY “WESTERN MOYAMOYA”. AND IT’S MORE FREQUENTLY SEEN, IN OUR EXPERIENCE, IN THE HEMORRHAGIC FORM OF MOYA MOYA DISEASE.

2- OUR EXPERIENCE SHOWS THAT THE MOST IMPORTANT POINTS RELATED WITH THE CLINICAL RESULTS ARE THE INITIAL NEUROLOGICAL DAMAGE, AND THE EARLY DIAGNOSIS AND TREATMENT.

3- WE EMPLOYED THE WIDELY USED TECHNIQUE OF EDAS, AND LATER, EDAS PLUS WIDER GALEAL FLAP, AND BURR HOLES, WITH GOOD ANGIOGRAPHICAL RESULTS AND RELATIVE GOOD CLINICAL RESULTS.
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THANK YOU VERY MUCH !!!