MULTIPLE BUR HOLES SURGERY FOR THE TREATMENT OF MOYAMOYA DISEASE IN CHILDREN

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• **Moyamoya syndrome**: a cerebrovascular condition, with *progressive stenosis* of the intracranial internal carotid arteries and their proximal branches

• Not associated conditions
  - Moyamoya disease
INTRODUCTION

• 1957, Takeuchi & Shimizu - Japan

• “something hazy, like a puff of cigarette smoke”
EPIDEMIOLOGY

• The high incidence: Japanese and Asian population

• Annual incidence: 0.35 to 0.94 per 100,000

• Prevalence: 3.2 to 10.5 per 100,000

• M:F = 1:1.8 to 1:2.2

• All ages: 1995 (10-14 yrs & 40-49 yrs)
  2008 (45-49 yrs & 5-9 yrs)
## Associated characteristics and conditions

<table>
<thead>
<tr>
<th>Common</th>
<th>50–75</th>
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<tbody>
<tr>
<td>Angiographic findings of moyamoya without other disease</td>
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<tr>
<td>Asian heritage</td>
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<tr>
<td><strong>Less common (moyamoya syndrome)</strong></td>
<td>10–20</td>
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<tr>
<td>Sickle cell disease</td>
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<tr>
<td>Neurofibromatosis type 1</td>
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<td>Cranial therapeutic irradiation</td>
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<tr>
<td>Down's syndrome</td>
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<td><strong>Rare (moyamoya syndrome)</strong></td>
<td>&lt;10</td>
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<tr>
<td>Congenital cardiac anomaly</td>
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<tr>
<td>Renal-artery stenosis</td>
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<tr>
<td>Giant cervicofacial hemangiomas</td>
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<td>Hyperthyroidism</td>
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Hyperproliferation of the vessel wall components and abundant intraluminal thrombi
In children, symptomatic episodes of ischemia may be triggered by exercise, crying, coughing, straining, fever, or hyperventilation.

<table>
<thead>
<tr>
<th>Symptoms at presentation</th>
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<tbody>
<tr>
<td><strong>Common</strong></td>
<td></td>
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<tr>
<td>Ischemic stroke</td>
<td>50–75</td>
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<tr>
<td>Transient ischemic attack (including drop attacks)</td>
<td>50–75</td>
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<tr>
<td>Hemorrhage (in adults)</td>
<td>10–40</td>
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<tr>
<td><strong>Less common</strong></td>
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<tr>
<td>Seizures</td>
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<tr>
<td>Headache</td>
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<tr>
<td><strong>Rare</strong></td>
<td></td>
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<tr>
<td>Choreiform movements</td>
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<td>Cognitive or psychiatric changes</td>
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IMAGING

• CT- CTA
• MRI- MRA
• DSA
• Tends to be progressive, bilateral:

Progressive neurologic deficits and poor outcome: 50 to 66 percent

Two thirds of patients with moyamoya have symptomatic progression over a 5-year period

More rapid progression and a worse prognosis in younger than in older children.

Prognosis: Poor without treatment
Medical treatment

Treatment

Endovascular Therapy

Surgery

Direct

STA-MCA bypass

Indirect

Encephalo… synangiosis

Multiple burr holes
1984, Masataka Endo –

Neovascularization occurring across a frontal burr hole / external drainage of intraventricle hemorrhage.
CASE REPORTS

Girl, 13 year-olds.
CASE REPORTS

- Girl, 13 month-olds

R

L
Multiple Bur Hole Surgery For The Treatment Of Moyamoya Disease In Children.

CHRISTIAN SAINTE-ROSE, M.D., RICARDO OLIVEIRA, M.D., STÉPHANIE PUGET, M.D., LIANA BENI-ADANI, M.D., NATHALIE BODDAERT, M.D., JOHN THORNE, F.R.C.S., ALISON WRAY, F.R.A.C.S., MICHEL ZERAH, M.D., AND MARIE BOURGEOIS, M.D

14 patients (mean age 6.5yr) – France & Australia

Post-op MRI: restoration of cortical perfusion as early as 3 months

Conclusion: effective and safe.

Furthermore, it is effective as a sole treatment without supplementary revascularization procedures.
Multiple Burr Hole Surgery As A Treatment Modality For Pediatric Moyamoya Disease

RAVINDRANATH KAPU, NIGEL PETER SYMSS, [...] AND RAVI RAMAMURTHI

- 10 year-old boy - India
- Bilateral multiple burr holes
- DSA 6 months post-op, excellent cerebral revascularization around the burr hole sites
- Conclusion: effective and safe, without supplementary revascularization procedures.
Effect Of Multiple Cranial Burr Hole Surgery On Prevention Of Recurrent Ischemic Attacks In Children With Moyamoya disease.

(http://www.ncbi.nlm.nih.gov/pubmed/20446218)

OLIVEIRA RS, AMATO MC, SIMÃO GN, ABUD DG, AVIDAGO EB, SPECIAN CM, MACHADO HR.

• Brasil – 7 pts
• No recurrence of ischemic attacks postoperatively.
• Neoangiogenesis was observed in both hemispheres
• Conclusion: simple procedure, relatively low risk of complications, effective for preventing cerebral ischemic attacks
Long Time Follow Up Of Patients With Moyamoya Disease Treated By Multiple Burr Hole Surgery

YOSHITO UCHIHASHI¹; KOHKICHI HOSODA¹; TETSURO KAWAGUCHI²; MITSUGU NAKAMURA³; EIJI KOHMURA

- Japan - 46 pts (mean age 40)
- Average Follow up period: 10.8 yrs
- Conclusion:
  - similar long-term results compared with direct bypass
  - easily done, performed in any place
Hemodynamic Change Of Childhood Moyamoya Disease After Multiple Burr-hole Operation

SUN JIAN*, WANG LI-SHU, ZHANG DONG, ET AL

• 8 pts – China (Chinese Journal of Stroke)
• Using multicolor ultraphonic power Doppler + SPECT
• 1st week post-op: neovascularization
• 3rd month: rich neovascularization.
• Conclusion: The burr-hole operation can improve the ischemia of childhood moyamoya disease
Conclusion: Burr hole surgery is an important and versatile tool in the armamentarium of surgeons treating children and adults with MMD, allowing tailoring of the revascularization techniques, or more recently alone in the setting of multiple burr holes. The superficial temporal artery and middle meningeal artery have both been found to contribute to revascularization via burr holes.

CONCLUSIONS: Burr hole surgery is an important and versatile tool in the armamentarium of surgeons treating children and adults with MMD, allowing tailoring of the revascularization. Further studies should help to determine factors that may help predict optimal revascularization from this surgical approach.
CONCLUSION

• Moyamoya disease- chronic progressive cerebrovascular disease- bilateral stenosis or occlusion of the arteries around the circle of Willis

• Patients often suffer cognitive and neurologic decline due to repeated ischemic stroke or hemorrhage

• Outcome: poor without treatment

• MBHO: effective and safe, and can be used as the only treatment without supplementary revascularization procedures

• In children, symptomatic episodes of ischemia may be triggered by exercise, crying, coughing, straining, fever, or hyperventilation
Thank You For Your Attention