

MOYAMOYA DISEASE IN HONG KONG: NATURAL HISTORY AND SURGICAL REVASCULARIZATION OUTCOME.

Anderson Chun On TSANG¹, Wai Man LUI¹, Yin Chung PO², Hok Ming CHIU³

¹Department of Neurosurgery, Queen Mary Hospital, Hong Kong

²Department of Neurosurgery, Princess Margaret Hospital, Hong Kong

³Department of Neurosurgery, Queen Elizabeth Hospital, Hong Kong

OBJECTIVE: Moya-moya disease (MMD) is an occlusive cerebral vasculopathy associated with high risk of recurrent ischemic and hemorrhagic events. Surgical revascularization had proven benefits in ischemic MMD but the role in hemorrhagic presentations is less clear. We studied the natural history of symptomatic MMD patients in Hong Kong and compared the long term outcome of revascularized patients with those treated conservatively.

METHODS: Patients with MMD managed in three high volume neurosurgical centers from 1997-2015 in Hong Kong were identified from the Hospital Authority's clinical data analysis and reporting system. The demographic and clinical characteristics, surgical outcome, and follow-up information were retrospectively reviewed. The primary outcome was recurrent hemorrhagic and ischemic stroke. The annual stroke risk and functional status of patients treated surgically and conservatively was compared.

RESULTS: Our cohort included 101 patients with a mean follow-up of 80.2 months. Female-to-male ratio was 1.8:1. 85.1% of patients had bilateral disease. The mean age of symptom onset was 42.6 years, with a bimodal distribution. 54.5% presented with intracranial hemorrhage and 39% with cerebral ischemic symptoms. 64 patients were treated conservatively, 15 underwent direct vascular bypass and 22 had indirect revascularization. 20 of the 64 (31.3%) patients treated conservatively had rebleeding, compared with 5 of 32 (15.6%) of surgically revascularized patients. The overall annual stroke risk per patient was significantly lower in revascularized than conservatively managed patients (4.6%/year vs 8.3%/year (p=.015)). 2 (5.4%) patients in the surgical group and 12 (18.8%) in the conservative group died of MMD during the follow up period. Surgically treated patients were more likely to have good functional outcome (mRS 0-2) at last follow up. (86.1% vs 62.5%, p=.024)

CONCLUSION: Our results confirmed the devastating natural history with high recurrent stroke risk in amongst Hong Kong MMD patients treated conservatively. Surgical revascularization conferred significant risk reduction for recurrent adverse neurological events, in particular that of hemorrhagic stroke. This translated to favourable survival and functional outcome compared with conservatively managed patients