Moyamoya Associated with Alagille Syndrome: A Multicenter Study of Surgical Revascularization and Outcomes

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Introduction

Seven high-volume centers in North America were surveyed to identify patients with coexistent moyamoya syndrome and Alagille syndrome (ALGS) (a multisystem disorder associated with abnormalities of the vasculature, liver, heart, skeleton, eyes and facies) in order to characterize the clinical and radiographic manifestations of moyamoya syndrome in this population, coupled with its response to indirect surgical revascularization.

Methods

Records from 7 centers were retrospectively reviewed. Patients were < 21 years of age, and met criteria for the diagnosis of both ALGS and moyamoya syndrome. All patients had been treated with indirect surgical revascularization, using variations of encephaloduralarteriosynangiosis, including pial synangiosis.

Results

Five patients with ALGS underwent revascularization. The majority of patients presented with ischemic symptoms, with radiographic or clinical stroke evident in 3 (60%) patients and transient ischemic attacks (TIA) in 4 (80%) at time of diagnosis. Radiographically, the arteriopathy was indistinguishable from moyamoya disease. The mean age at surgery was 2.5 [1.7-4] years with one female and 4 males. Complications included 1 patient who suffered a stroke within 30 days following surgery. No late-onset strokes occurred. Mean follow-up period was 7.4 (1-13) years. Clinically, all patients had a modified Rankin Score of 0-1, with stable or improved neurologic examinations. Radiographically, all 5 patients have evidence of revascularization on neuroimaging and no subsequent strokes.

Conclusions

Clinical and radiographic features of moyamoya syndrome associated with ALGS are comparable to primary moyamoya. Indirect surgical revascularization is safe in Alagille-associated moyamoya and confers demonstrable long-term clinical and radiographic protection against stroke.

Learning Objectives

By the conclusion of this session, participants should be able to: 1) Describe the importance of surgical revascularization in patients with moyamoya associated with Alagille Syndrome, 2) Assess the importance of screening for moyamoya in patients who carry a diagnosis of Alagille syndrome.

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