Rapid Clinical/Radiographic Deterioration in Patients with Moyamoya Disease

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Author report 20 patients who have shown rapid clinical and/or radiographic deterioration among 168 patients (Pts) (48 children and 120 adults) who underwent an extracranial to intracranial bypass surgery for moyamoya disease (MMD).

They included 12 child-onset MMD and 8 adult-onset MMD patients with age ranging from 17 months to 37 years.

Results: 17 Pts (85%) of 20 Pts presented with cerebral ischemia or infarction related symptoms or signs. 18 Pts underwent combined direct/indirect or multiple indirect bypass surgery prior to their deterioration (unilateral in 17 Pts and bilateral in one Pt).

Two pediatric Pts (17-month-old and 24-month-old) showed a fulminant clinical course associated with bilateral radiographic progression despite they underwent bilateral bypass surgery.

16 Pts (80%) of 20 Pts progressed within 10 months after an initial study clinically or radiographically regardless of the site or the method of bypass surgery.

7 Pts (4 children and 3 adults) demonstrated clinical deterioration accompanied with the radiographic progression.

Conclusion: These findings indicate that MMD can be progressed rapidly clinically and/or radiographically in pediatric Pts as well as adult Pts, although their frequencies are much lower in adult Pts as compared with pediatric Pts.

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