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1: [J Child Neurol](#). 2000 Aug;15(8):559-62.

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## Intracranial fibromuscular dysplasia in a six-year-old child: a rare cause of childhood stroke.

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Intracranial fibromuscular dysplasia is a nonatheromatous angiopathy that most commonly affects adult women and is rarely recognized in children. Symptoms include stroke and headache, although the vasculopathy may be asymptomatic. Diagnosis is based on angiographic appearance, commonly described as a "string of beads." The etiology of intracranial fibromuscular dysplasia is not known, although possible causes include genetic predisposition, trauma, and underlying connective tissue disease. Treatment of intracranial fibromuscular dysplasia is largely supportive once symptoms become manifest. We report a 6-year-old girl who presented to our center for further evaluation of a large left middle cerebral artery distribution infarction. The patient was previously healthy, without known risk factors for stroke. Initial symptoms consisted of a dense global aphasia and a right hemiparesis. On arrival, the patient's aphasia had improved but she continued to have significant deficits in both receptive and expressive language as well as residual right hemiparesis. Magnetic resonance imaging and conventional angiographic studies demonstrated characteristic beading of the distal portion of the left internal carotid artery, as well as the proximal middle cerebral artery. Laboratory evaluation, echocardiogram, and renal ultrasound were normal. The renal vasculature did not demonstrate evidence of intracranial fibromuscular dysplasia. In conclusion, intracranial fibromuscular dysplasia should be considered in the differential diagnosis of childhood stroke. When recognized, other sites of vascular involvement should be sought, and consideration of underlying disorders is important, as connective tissue disorders have been associated with a propensity to develop this vascular abnormality. Careful follow-up is warranted, due to possible progression of disease.

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PMID: 10961798 [PubMed - indexed for MEDLINE]

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