"Moyamoya disease is a progressive disorder that affects the blood vessels in the brain (cerebrovascular). It is characterized by the narrowing (stenosis) and/or closing (occlusion) of the carotid artery inside the skull, a major artery that delivers blood to the brain. At the same time, tiny blood vessels at the base of the brain open up in an apparent attempt to supply blood to the brain distal to the blockage. These tiny vessels are the ""moyamoya"" vessels for which the disease was named. Inadequate blood supply then leads to reduced oxygen delivery to the brain, and it is this oxygen deprivation that causes the signs of moyamoya. One of the symptoms is typically stroke, which results in paralysis of the face, arms or legs, loss of speech, etc., or temporary loss of neurologic function of body parts or speech (transient ischemic attacks, or “TIA”). Other symptoms that may result include headaches, visual disturbances, developmental delay, and seizures. Approximately 10% of cases of moyamoya in Asian countries have a genetic cause. Patients with this arteriopathy that occurs either on a familial or idiopathic basis are said to have moyamoya disease. Patients in whom the artery changes occur in association with another process such as sickle cell disease or Down syndrome are said to have moyamoya disease. In this report, we use the term ""moyamoya disease"" as shorthand for both forms. In Japan, moyamoya disease typically occurs in females under the age of 20. In Japan, the disease is estimated to occur in 1 per 300,000 people. Although moyamoya was originally reported in individuals of Japanese ancestry, cases have been reported from elsewhere in Asia as well as from Europe, North and South America, and most series reported in the western hemisphere have a minority of patients of Asian descent. Of note, most patients in North America are isolated cases, with recent literature suggesting that less than 4% of cases in this population are familial. (Gaillard 2017) In most patients, the diagnosis of moyamoya can be made from a careful assessment of an MRI and MRA. Cerebral arteriography will confirm the diagnosis, establish the exact degree of blood vessel narrowing, demonstrate the existing blood flow patterns to various areas of the brain, and allow treatment decisions to be made; for these reasons, it is the standard diagnostic tool for this condition. In particular, catheter angiography can help with the identification of important blood vessels called “transdural collaterals,” which are present in some cases and can markedly influence surgical planning and prognosis. (Storey 2017)"