

Fibromuscular dysplasia, commonly called FMD, is a disease that causes one or more arteries in the body to have abnormal cell development in the artery wall. As a result, areas of narrowing (stenosis), aneurysms, or tears (dissection) may occur. If narrowing or a tear causes a decrease in blood flow through the artery, symptoms may result. FMD is most commonly found in the arteries that supply the kidneys with blood (renal arteries) and the arteries called the carotid and vertebral arteries which are found in the neck and supply the brain with blood. Less commonly, FMD affects the arteries in the abdomen (supplying the liver, spleen and intestines) and extremities (legs and arms). In more than one-half of people with this disease, there will be evidence of FMD in more than one artery. FMD affects women far more commonly than men, although men and children can be affected with this disease. In children with FMD, the disease seems to more commonly present with significant narrowing rather than tears of arteries and also seems to involve the arteries to the kidneys and intestines more commonly than the carotid vessels. In the pediatric population, FMD affects both boys and girls. The vascular subtype of Ehlers-Danlos syndrome (type IV) has been associated with the most common type of fibromuscular dysplasia, known as multifocal FMD. This syndrome should be suspected in patients with multiple aneurysms and/or tears (dissections) in arteries in addition to the typical angiographic findings of fibromuscular dysplasia. There have been isolated reports of fibromuscular dysplasia associated with other disorders, including Alport syndrome, pheochromocytoma, Marfan syndrome, and moyamoya disease. (For more information on these disorders, search for the term in the Rare Disease Database.) In order to diagnosis FMD, a test must be done to image the blood vessels. There are many options for imaging the arteries, including specialized blood vessel ultrasound known as duplex ultrasound; a CAT scan of the arteries which is obtained after a dye is given through the veins, or a special type of MRI. In many cases, the diagnosis of FMD requires that a procedure known as an arteriogram be performed. Arteriography is a procedure that is performed by a radiologist, vascular surgeon, cardiologist, or vascular medicine specialist with appropriate training. It involves inserting a wire into or near the affected artery and injecting contrast material, a dye that can be detected by an X-ray machine. An X-ray of the affected area is then taken and examined. The individual is usually awake during an arteriogram procedure although medications may be given to keep her or him comfortable. This outpatient procedure usually lasts from one to two hours with a recovery period of up to six hours (this varies widely).