Standard Therapies

There is no cure for FMD. Treatments are focused on managing symptoms and complications of FMD, including high blood pressure and headaches. Antiplatelet medications, such as aspirin, may be prescribed along with medications to treat high blood pressure (anti-hypertensives). A number of medications are available to help control and prevent headaches. All patients with FMD who use tobacco should be encouraged to quit.

In some cases of FMD, an attempt should be made to improve the flow of blood through a severely narrowed vessel. The kind of treatment used for narrowing due to FMD depends largely upon which arteries are affected and the presence and severity of the symptoms.

In most cases, such procedures are done using balloon angioplasty, a procedure known as percutaneous transluminal angioplasty (PTA). PTA is often performed at the same time as an arteriogram. If an angioplasty is performed, a catheter is extended into the affected artery and a small balloon is inflated in the artery. A metal stent is typically not required to keep the vessel open, but may be needed in some cases, such as for treatment of a tear (dissection) of a blood vessel. If angioplasty is performed, the procedure and recovery period may be longer than an arteriogram done for diagnosis only.

Occasionally, open surgery is performed to treat severe narrowing due to FMD, particularly those that cannot be treated with angioplasty.

Patients with FMD who are found to have a significant aneurysm within the brain or renal arteries may need to undergo surgery even if they do not have any symptoms. In such cases, it is recommended that the aneurysm be treated to prevent rupture, which can be potentially life threatening. The type of treatment for an arterial aneurysm depends on its location and size. Treatment options include open surgery or a less invasive angiogram-based procedure, which treats the aneurysm using special vascular coils and/or stents.

The appropriate treatment will vary with each individual and severity, location, and extent of disease. The treatment plan should be discussed in depth with a specialist who is knowledgeable about FMD and its natural history.

Research and Investigational Therapies

The FMD Society of America is sponsoring the United States patient registry for FMD, which is gathering information regarding the epidemiology, symptoms, disease extent, treatments, and outcomes of patients who have FMD. Currently we have 13 Centers participating in the Patient Registry and to date almost 1400 patients have participated. To learn more about the FMD patient registry, go to www.fmdsa.org/research_network/fmd_registry

Updated October 2015, Pamela Mace RN, Executive Director FMDSA and Dr. Heather Gornik, UH Cleveland Medical Center and FMDSA Medical Advisory Board.

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FIBROMUSCULAR DYSPLASIA SOCIETY OF AMERICA

Fibromuscular Dysplasia

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.

Types of Fibromuscular Dysplasia

The most common type of fibromuscular dysplasia is an artery that has the appearance of a string of beads on imaging; this is referred to as multifocal FMD. Focal FMD is less common, has the appearance of a focal or tubular stenosis, and it is most commonly diagnosed in children. Some patients can have both multifocal and focal FMD.

Symptoms

The signs and/or symptoms that a person with FMD may experience depend on the arteries affected and whether there is narrowing, tears, or aneurysms within them. Some patients with FMD may have no symptoms but are diagnosed with this disease when a physician hears a noise over one of the arteries due to disturbed or turbulent blood flow within the vessel (known as a bruit), or when they have imaging studies done for other reasons.

Any pain or clinical sign related to FMD typically comes from the organ that is supplied by that artery. For example, FMD in the kidney arteries may cause high blood pressure. FMD in the carotid arteries may cause headaches or a swooshing sound in the ears (pulsatile tinnitus). FMD involving the arteries that supply the intestines, liver and spleen with blood (mesenteric arteries) can result in abdominal pain after eating and unintended weight loss. FMD in the arms and legs can cause limb discomfort with exercise or can lead to unequal blood pressures in the arms. A person with carotid FMD causing severe narrowing or a tear in a carotid or vertebral artery may have neurologic symptoms involving the facial nerves (such as drooping of the eye lid, unequal size of the pupils), stroke or transient ischemic attack. People with carotid FMD have a higher risk for aneurysms of the arteries in the brain (intracranial aneurysms). Bleeding in the brain (intracranial hemorrhage) may occur if an aneurysm ruptures, and it is important to identify and treat brain aneurysms early to prevent this.

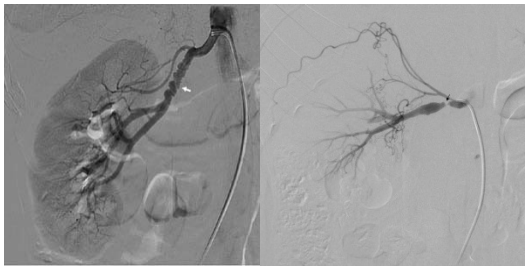
Recently, studies have shown that some women who were healthy and who have a sudden tearing of a coronary artery (called Spontaneous Coronary Artery Dissection or "SCAD") may have undiagnosed FMD. Previously, SCAD was thought to be a separate condition from FMD, but now we are starting to understand that many people have FMD as the main reason for their coronary artery tear. It is important to note, however, that the majority of patients who have FMD and are seen in follow-up never will have SCAD.

Causes

The cause of FMD is not yet known, but several theories have been suggested. A number of case reports in the literature have identified the disease in multiple members of the same family including twins. As a result, it is felt that there may be a genetic cause. However, a relative may have different artery involved, different disease severity, or may not develop FMD at all. In fact, most individuals with FMD do not have a family member who also has the disease. Among some individuals with FMD, there is a family history of other vascular problems, such as blood vessel aneurysms.

FMD is far more commonly seen in women than in men, resulting in the theory that hormones may play an important role in disease development. However, in small population studies, one's reproductive history (number of pregnancies and when they occurred) as well as use of birth control pills did not correlate with the development of FMD.

Other possible causes of FMD include abnormal development of the arteries that supply the vessel wall with blood, resulting in inadequate oxygen supply; the anatomic position or movement of the artery within the body; certain medications, and tobacco use. It is possible that many factors contribute to the development of FMD. This area requires further research.



Multifocal FMD of the renal artery (left); focal FMD (right)

Affected Populations

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 presents 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.

Related Disorders

The vascular sub type of Ehlers-Danlos syndrome (type IV) has been associated with the most common type of fibromuscular dysplasia, known as medial fibroplasia. 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's syndrome, pheochromocytoma, Marfan's syndrome, and moyamoya disease.

Diagnosis

In order to diagnosis FMD, a test must be done to image the blood vessels. Options include a specialized blood vessel ultrasound known as duplex ultrasound, a CT 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. 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).