

Fibromuscular Dysplasia

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The circulatory system is made up of the heart and blood vessels. There are 3 major types of blood vessels: arteries, veins, and lymphatics. The heart is the pumping organ and helps to push oxygen- and nutrient-rich blood through the arteries to the organs and the limbs. The veins return blood from the organs and limbs to the heart so that it can be resupplied with oxygen from the lungs. The lymphatics connect with the veins and help to return fluid from tissues and skin. This article focuses on a disorder of the arteries known as fibromuscular dysplasia.

What Is

Fibromuscular Dysplasia?

Fibromuscular dysplasia (also known as FMD) is a medical condition characterized by abnormal cell growth in the walls of arteries of the body. FMD is different from other blood vessel disorders that affect the arteries, such as atherosclerosis (blockage of arteries secondary to cholesterol plaque), vasculitis (inflammation of the arteries), and thrombosis (formation of blood clots). FMD can lead to different abnormalities of arteries,

such as narrowing, a beaded appearance (Figure, A and B), or even serious complications such as aneurysms (bulging of the arteries) or dissections (tears of the arteries). FMD is not known to involve the veins or the lymphatic system.

FMD most commonly affects the arteries that supply blood to the kidneys (renal arteries) and brain (carotid and vertebral arteries), but it can occur in almost any artery, including those that supply the intestines (mesenteric arteries), the arms (brachial arteries), and the legs (iliac arteries). In many patients, FMD is found in more than 1 artery.

FMD is classified according to the layer of the artery wall that is most involved (Figure). The most common type is medial fibroplasia. Medial fibroplasia affects the middle layer of the artery wall and leads to a very typical beaded appearance of the vessel, which some call a string of beads. Intimal fibroplasia is a less common type that affects the inner layer of the artery wall and leads to an area of smooth narrowing of the vessel.

Who Is Affected by FMD?

FMD is considered uncommon, but the frequency of this condition in the

general population is not known. Recent studies suggest that it is not as rare as initially thought. The majority of patients diagnosed with FMD are female; however, men can also have FMD. Data from the United States FMD patient registry suggest that patients are typically diagnosed with FMD in their early 50s and that there is no significant difference in the age of diagnosis between men and women. A percentage of patients with FMD are diagnosed in childhood. In children, intimal fibroplasia is the most common type.

What Causes FMD?

The cause of FMD is unknown. A number of theories have been proposed, including environmental factors, such as smoking and estrogen, as well as genetic factors. Approximately 10% of patients with FMD have an affected family member. Understanding the cause(s) of FMD continues to be an area of active investigation.

What Are the Symptoms and Signs of FMD?

Patients with FMD experience a variety of symptoms that are largely de-

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(*Circulation*. 2012;125:e636-e639.)

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Circulation is available at <http://circ.ahajournals.org>

DOI: 10.1161/CIRCULATIONAHA.111.090449

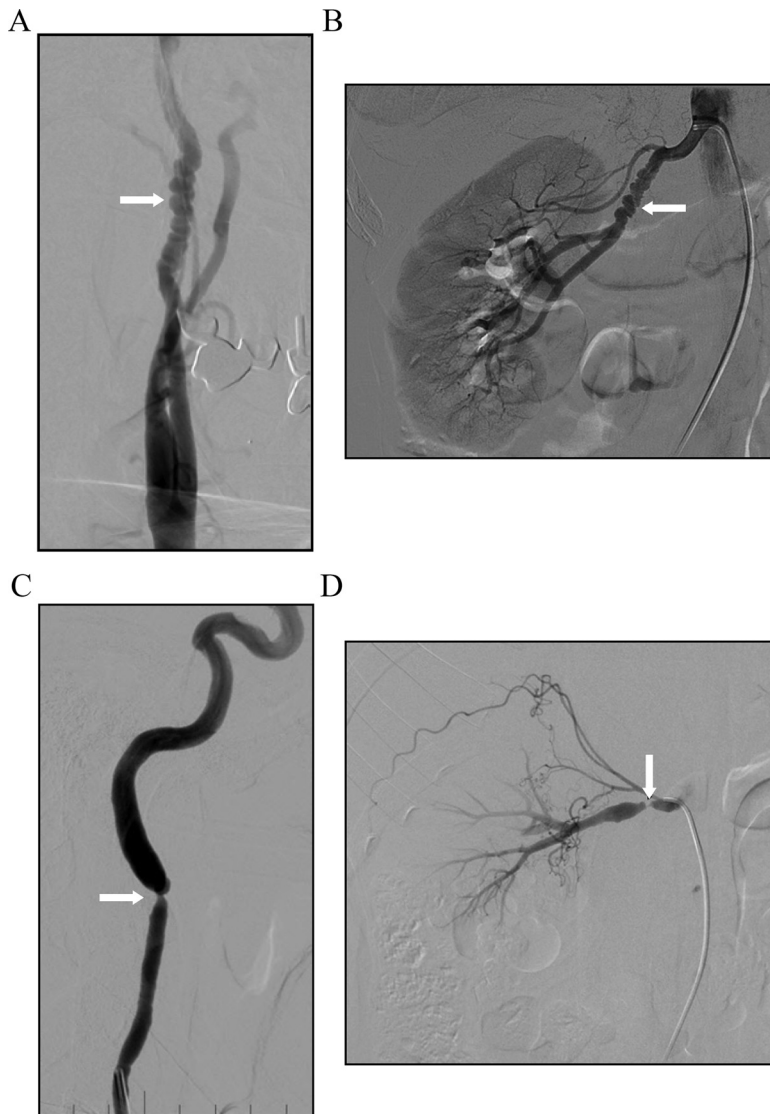


Figure. Fibromuscular dysplasia in the carotid and renal arteries as imaged with angiography, the gold standard test. **A** and **B**, Medial fibroplasia in the internal carotid (**A**) and renal (**B**) arteries with the classic string of beads appearance. **C** and **D**, The less common intimal fibroplasia, which presents as a focal, bandlike narrowing in the internal carotid (**C**) and renal (**D**) arteries.

pendent on the location of the affected arteries (Table); however, many patients have no symptoms at all and are diagnosed when an imaging test of the arteries is performed for another reason. Patients with carotid artery FMD may experience headaches (especially migraine type), neck pain, and a pulsatile ringing or swooshing sound in the ears. When patients with carotid FMD are examined, physicians may hear a bruit in the neck (abnormal sound heard when listening over the

carotid artery) or may note abnormalities in the pupils of the eyes or the eyelids. The most common sign of renal artery FMD is high blood pressure. In some cases, a physician may hear a bruit when listening over the abdomen or the flanks. Patients with mesenteric (intestinal) FMD can have abdominal pain after eating or weight loss. If FMD involves the arteries of the arms or legs, patients can experience pain with exertion that is relieved by rest, also known as claudication.

Table. Signs and Symptoms of FMD, Which Vary Depending on the Blood Vessels That Are Involved

Renal
High blood pressure
Bruit heard over the abdomen or flanks
Renal artery aneurysm
Renal artery dissection
Cerebrovascular
Headaches (especially migraine type)
Pulsatile ringing or swooshing noise in ears
Stroke or ministroke (TIA)
Neck pain
Dizziness
Bruit heard over the carotid arteries
Brain aneurysm
Carotid or vertebral artery dissection
Other sites
Abdominal pain after eating (mesenteric FMD)
Weight loss (mesenteric FMD)
Arm or leg pain with exertion (lower or upper extremity FMD)
Femoral bruit (iliac FMD)
Heart attack caused by coronary artery dissection (rare)

FMD indicates fibromuscular dysplasia; TIA, transient ischemic attack.

What Are Warning Signs of FMD?

Patients with FMD can develop an aneurysm or dissection in the affected arteries that can cause severe pain. Depending on the artery involved, this may lead to new-onset or worsening headache, neck pain, or abdominal pain. Unfortunately, some patients with carotid artery FMD experience stroke. Patients with FMD and sudden changes in vision or speech, new-onset weakness in an arm or leg, and/or alterations in consciousness should seek immediate medical care.

How Is FMD Diagnosed?

All patients in whom a diagnosis of FMD is being considered should undergo a detailed medical history and thorough vascular examination; however, a definitive diagnosis of FMD can only be made with imaging studies. Catheter-based angiography is the most accurate imaging technique. Dur-

ing the angiography procedure, a catheter (long slender tube) is inserted into a large artery and slowly advanced until it reaches the vessel of interest. A small amount of contrast is then injected through the catheter, and radiographic (x-ray) pictures of the arteries are taken. The use of contrast material is necessary for the arteries to appear on the radiographic pictures. Catheters may also be used to measure the pressure inside of an artery, which helps to determine the severity of disease. Computed tomography angiography and magnetic resonance angiography are other imaging techniques that may be used to diagnose FMD. These are particularly useful to evaluate arteries in the brain. Doppler ultrasound may be used in both the diagnosis and follow-up of FMD. Note that computed tomography angiography, magnetic resonance angiography, and ultrasound are noninvasive imaging techniques; therefore, no catheter is placed inside the body to image the arteries when these are performed.

What Treatment Options Are Available?

There is no cure for FMD. Many patients with carotid or vertebral artery involvement will be prescribed aspirin to reduce the risk of stroke. Patients with renal artery FMD often take medications to help control the blood pressure, particularly certain classes of blood pressure medications known as angiotensin-converting enzyme inhibitors or angiotensin receptor blockers.

Some patients with FMD may be advised to undergo percutaneous balloon angioplasty. The goal of balloon angioplasty is to widen a narrowing in the affected artery to improve blood flow. Improved blood flow in the renal arteries may result in better blood pressure control, whereas improved blood flow in the carotid arteries may help to prevent stroke. During angioplasty, a balloon on a guidewire is inserted through a catheter (tube) placed in an artery in the groin and advanced to the artery that is narrowed. The balloon is

then inflated to open the artery, deflated, and withdrawn.

Angioplasty is recommended for patients with renal artery FMD who have uncontrollable blood pressure, intolerance of medications, or declining kidney function. For some patients with newly diagnosed high blood pressure caused by renal artery FMD, angioplasty may also be recommended. Unlike patients with renal artery disease caused by atherosclerosis (or plaque), for patients with renal FMD, balloon angioplasty is generally performed without placement of a metallic stent. Angioplasty is most likely to be effective at controlling or curing high blood pressure in patients who do not have other risk factors associated with high blood pressure (such as obesity or diabetes mellitus) and in those who have had high blood pressure for a short period of time (<5 years).

In some cases, angioplasty may also be performed in patients with carotid or vertebral artery FMD who have severe neurological symptoms. For symptomatic patients with carotid or vertebral artery FMD who have suffered a dissection (tear), angioplasty with placement of a metallic stent may be performed. In less common cases, patients with FMD in the arteries of the arms, legs, or intestines may also undergo an angioplasty procedure to relieve symptoms caused by the arterial narrowing.

Patients with FMD who have a significant aneurysm of an artery may be referred for a procedure even if there are no symptoms. Aneurysms in the intracranial (brain) arteries can be treated with open surgery or placement of coils and special stents to seal off the aneurysm sac, a procedure that is performed from a catheter placed in an artery in the groin. Patients with large renal aneurysms may require surgical repair if these cannot be treated with endovascular therapies (coils and stents).

Are There Activity Restrictions for Patients With FMD?

Advice regarding physical activity should always be customized to each

individual patient and depends on factors such as vascular location and severity of disease, history of artery dissection or aneurysm, and blood pressure control. In general, experts recommend against contact sports, skydiving, scuba diving, and heavy weight lifting for patients with carotid FMD, prior dissection, or aneurysms, because these put added stress on arteries that are already prone to injury. Because of sudden jerking motions of the head, most advise against roller coaster or thrill rides at the amusement park and chiropractic manipulation or deep tissue massage of the neck for patients with carotid artery FMD. Although regular aerobic exercise (such as walking or cycling) is an important part of vascular health for FMD patients, every patient should discuss exercise plans with a physician who is familiar with FMD.

Who Cares for Patients With FMD?

FMD is a chronic medical condition and requires periodic visits with healthcare providers who are familiar with this problem. Healthcare providers who may be trained in the care of FMD patients include cardiologists, interventional radiologists, nephrologists, neurologists or neurosurgeons, vascular medicine specialists, and vascular surgeons. In general, the experience of a provider in caring for patients with FMD is more important than their particular medical specialty. The FMD Society of America maintains a list of clinical experts experienced in the care of FMD patients (<http://www.fmdsa.org/>).

What Type of Follow-Up Care Is Recommended?

Depending on symptoms and clinical signs (such as high blood pressure or headaches), patients diagnosed with renal artery FMD may be advised to undergo an imaging study to test for involvement of the carotid arteries. Those with carotid artery disease may be advised to undergo imaging of the renal arteries. It is recommended that



patients with carotid FMD undergo an imaging test (such as magnetic resonance angiography) at least once to rule out brain aneurysms. Imaging of other blood vessels (such as the arteries in the arms, legs, and intestines) is typically performed only in those with signs or symptoms of disease.

For FMD patients with stable symptoms, periodic clinical follow-up visits will focus on symptom control, blood pressure, and the vascular examination. Imaging studies are performed periodically to assess for progression of disease. Some patients with stable FMD will not need to be seen more frequently than yearly. In contrast, follow-up should take place more often for those with severe or worsening symptoms. The type of imaging study used for follow-up of FMD (eg, magnetic resonance angiography versus ul-

trasound) depends on the arteries that are affected, whether the arteries can be well imaged with the ultrasound probe, and the particular imaging expertise of the facility.

What Resources Are Available for Patients With FMD?

The FMD Society of America (FMDSA) is a nonprofit organization committed to increasing awareness, education, and research related to FMD. The FMDSA sponsors an annual meeting for patients and health-care providers that is held each spring in Cleveland, Ohio, and maintains a Web site with up-to-date information (<http://www.fmdsa.org>). With sponsorship from the FMDSA, 10 health-care centers are currently enrolling patients in the United States FMD patient

registry (see Additional Resources). The FMD registry will help health-care providers and patients to better understand the clinical features and natural course of this disease. To date, >500 patients have consented to participate in the FMD patient registry.

Additional Resources

A list of clinical centers participating in the United States FMD patient registry is available at http://www.fmdsa.org/patient_support/patient_registry (accessed January 10, 2012).

Disclosures

Drs Gornik and Olin are volunteer (non-compensated) medical advisory board members to the FMDSA. P. Mace is a paid employee of FMDSA. S. Poloskey reports no conflicts.

KEY WORDS: fibromuscular dysplasia ■ patient education ■ peripheral vascular disease

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Circulation. 2012;125:e636-e639

doi: 10.1161/CIRCULATIONAHA.111.090449

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:

<http://circ.ahajournals.org/content/125/18/e636>

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