

Dissection and Aneurysm in Patients With Fibromuscular Dysplasia



Findings From the U.S. Registry for FMD

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ABSTRACT

BACKGROUND Fibromuscular dysplasia (FMD) is a noninflammatory arterial disease that predominantly affects women. The arterial manifestations may include beading, stenosis, aneurysm, dissection, or tortuosity.

OBJECTIVES This study compared the frequency, location, and outcomes of FMD patients with aneurysm and/or dissection to those of patients without.

METHODS The U.S. Registry for FMD involves 12 clinical centers. This analysis included clinical history, diagnostic, and therapeutic procedure results for 921 FMD patients enrolled in the registry as of October 17, 2014.

RESULTS Aneurysm occurred in 200 patients (21.7%) and dissection in 237 patients (25.7%); in total, 384 patients (41.7%) had an aneurysm and/or a dissection by the time of FMD diagnosis. The extracranial carotid, renal, and intracranial arteries were the most common sites of aneurysm; dissection most often occurred in the extracranial carotid, vertebral, renal, and coronary arteries. FMD patients with dissection were younger at presentation (48.4 vs. 53.5 years of age, respectively; $p < 0.0001$) and experienced more neurological symptoms and other end-organ ischemic events than those without dissection. One-third of aneurysm patients (63 of 200) underwent therapeutic intervention for aneurysm repair.

CONCLUSIONS Patients with FMD have a high prevalence of aneurysm and/or dissection prior to or at the time of FMD diagnosis. Patients with dissection were more likely to experience ischemic events, and a significant number of patients with dissection or aneurysm underwent therapeutic procedures for these vascular events. Because of the high prevalence and associated morbidity in patients with FMD who have an aneurysm and/or dissection, it is recommended that every patient with FMD undergo one-time cross-sectional imaging from head to pelvis with computed tomographic angiography or magnetic resonance angiography. (J Am Coll Cardiol 2016;68:176–85) © 2016 by the American College of Cardiology Foundation.



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Fibromuscular dysplasia (FMD) is a noninflammatory arterial disease that affects predominantly women. Arterial manifestations include beading, stenosis, aneurysm, dissection, and arterial tortuosity (**Central Illustration**) (1,2). The most common histological type of FMD is medial fibroplasia, which results in an artery with a “string of beads” appearance, representing alternating areas of stenosis due to fibrous webs and post-stenotic dilation. Due to the increasing use of endovascular therapy, tissue samples are rarely obtained in the current era; thus, it has been recommended that an angiographic classification scheme replace the previously used histopathological classification (3,4). The

string-of-beads type is now called “multifocal” FMD (3,4) (**Figure 1A**), whereas “focal” FMD denotes a single area of concentric or tubular stenosis, most commonly due to intimal fibroplasia, pathologically (**Figure 1B**) (3,5,6).

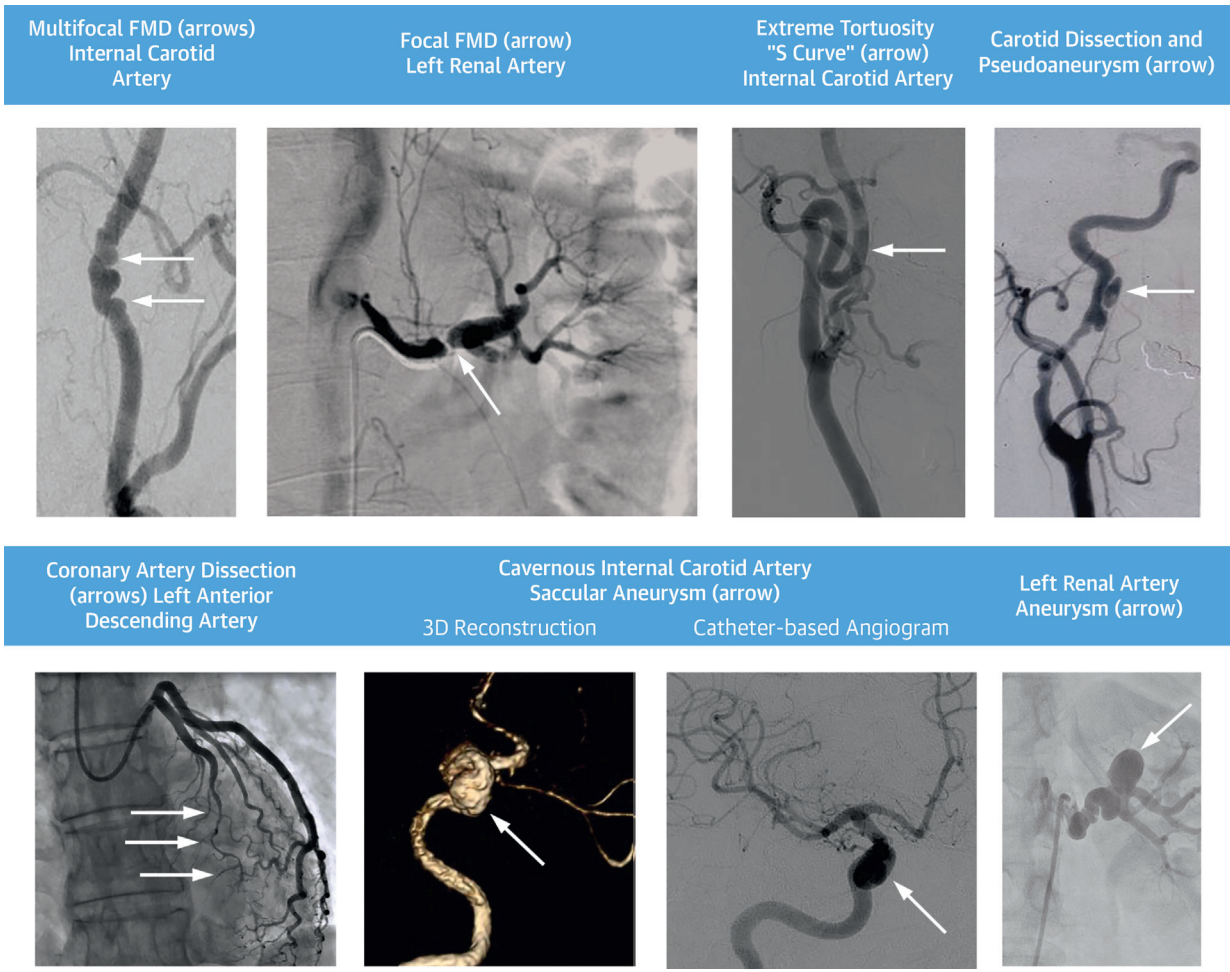
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The prevalence of FMD remains unknown; however, data from asymptomatic kidney donors suggest that up to 4% of the general population may have FMD (1,7). Prevalence may be even higher in patients with resistant hypertension (8). FMD affects predominantly women 20 to

**ABBREVIATIONS
 AND ACRONYMS**

- CTA** = computed tomographic angiography
- FMD** = fibromuscular dysplasia
- MRA** = magnetic resonance angiography
- SAH** = subarachnoid hemorrhage
- SCAD** = spontaneous coronary artery dissection
- TIA** = transient ischemic attack

CENTRAL ILLUSTRATION The Many Faces of Fibromuscular Dysplasia



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Imaging characteristics of FMD include a “string of beads” appearance to the artery, a focal area of stenosis, extreme tortuosity, dissection of a coronary or peripheral artery, and/or aneurysm of a peripheral artery, an intracranial artery, or the aorta. 3D = 3-dimensional; FMD = fibromuscular dysplasia.

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