

Treatment of Renal Artery Fibromuscular Dysplasia

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Fibromuscular dysplasia is a nonatherosclerotic and noninflammatory disease that can result in stenoses of the renal arteries and hypertension, most commonly affecting middle-aged women. Percutaneous transluminal angioplasty has long been considered the mainstay of therapy and offers high rates of improved or cured hypertension. The disease involves the mid and distal renal arteries and branchpoints and poses endovascular treatment challenges that separate fibromuscular dysplasia from atherosclerotic disease. The development of smaller balloon dilation systems offers safe and highly effective endovascular treatment options for technically difficult lesions. Newer technologies such as cutting balloons also add to the armamentarium of treatment choices, which may be useful in the setting of resistant stenoses. This article focuses on the modern technical considerations in the diagnostic evaluation and endovascular treatment of renal artery fibromuscular dysplasia.

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Fibromuscular dysplasia (FMD) is a nonatherosclerotic and noninflammatory disease that most commonly affects the renal and carotid arteries but can involve any artery.¹ The prevalence of FMD is approximately 0.5% to 5% of the population, and the disease has been gaining popularity in the recent press because of its potential underdiagnosis.² FMD occurs predominantly in middle-aged Caucasian women, and its pathogenesis is not fully understood. Genetic ties have been discovered between first-degree relatives, with the disease transmitted as autosomal dominant with incomplete penetrance and variable expression. Many cases have no family history, and risk factors, such as smoking and oral contraceptives, have also been associated. Even mechanical stress in the setting of renal ptosis has been theorized as a potential etiology.³

Histopathological classification of renal artery FMD as originally proposed by McCormack and coworkers is based on the predominant arterial wall layer involved.⁴ The three most common and classically described subtypes include medial (70% of cases), perimedial (15-25%), and intimal fibrodysplasias (1-2%).⁵ Both the medial and perimedial fibrodysplasias form alternating stenoses and aneurysms such that their angiographic

appearance is that of a “string of beads,” frequently involving the bilateral renal arteries. Medial fibrodysplasia tends to create aneurysms larger than the expected luminal diameter of the artery from which they originate and rarely leads to occlusion (Fig. 1). Perimedial fibrodysplasia creates aneurysms smaller than the expected luminal diameter, resulting in highly stenotic lesions than can progress to occlusion (Fig. 2). Intimal fibrodysplasia creates smooth narrowings that can be focal or tubular, typically occurs in children, and can progress to occlusions.⁶ Less common subtypes continue to be described, and any of the subtypes can coexist within a single patient (Fig. 3).

Percutaneous transluminal renal angioplasty (PTRA) has been the mainstay of treatment for renal FMD, with technical success rates approaching 100% and excellent reported rates of improved or even cured hypertension.⁷ According to recent American College of Cardiology guidelines, the indications for intervention include resistant hypertension, intolerance to hypertensive medications, noncompliance with medications, and renal impairment or cortical loss from ischemia.⁸ This article focuses on modern technical considerations in the treatment of renal FMD.

Patients presenting for treatment of FMD are typically younger, and therefore common femoral artery access is not usually complicated by calcific atherosclerotic changes. A diagnostic aortogram is part of the initial diagnostic workup, but alone does not provide adequate evaluation of the renal

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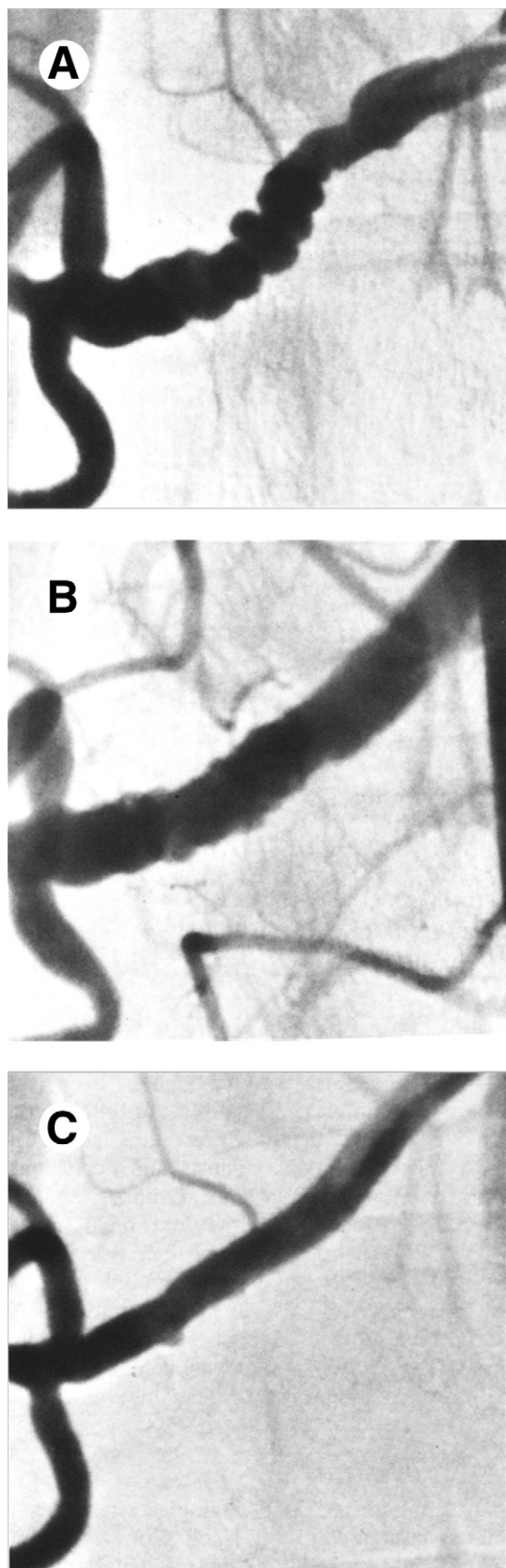


Figure 1 (A) In the medial fibroplasia type of FMD aneurysms are larger than the expected luminal diameter. (B) Immediate post-treatment image shows destruction of the web-like stenosis. The lumen is patent with some residual wall irregularity. (C) At 12 months, the lumen has healed to a more narrow caliber than the expected normal caliber of the vessel. (Images reprinted with permission.¹⁵)

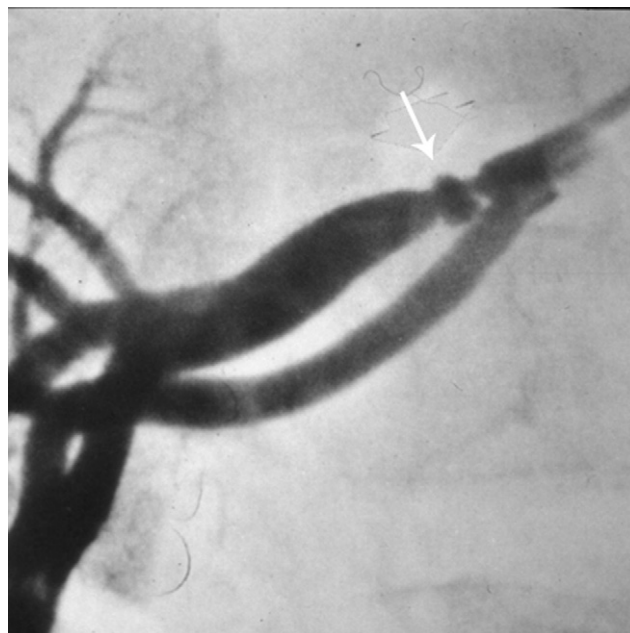


Figure 2 In the perimedial type of FMD aneurysms are smaller than the expected luminal diameter. (Image reprinted with permission.¹⁵)

arteries. Selective renal angiograms are essential, and often multiple views are required to fully profile the renal arteries and define the branch and intrarenal arterial anatomy. Digital subtraction angiography is the gold standard in diagnosis of FMD. Although the disease is detectable on ultrasound and magnetic resonance imaging, multidetector computed tomography angiography holds the greatest accuracy in the noninvasive diagnosis of FMD, with sensitivity and specificity reported to be 64-99% and 89-98%, respectively (Fig. 4).⁹

The diagnostic evaluation and interventional treatment of renal FMD can nearly always be accomplished via the common femoral artery. A diagnostic aortogram using a flush catheter and selective renal arteriograms using a reverse curve or downward-directed selective catheter (Sos II, Cobra 2, AngioDynamics, Glen Falls, NY; Simmonds I, Cook, Inc, Bloomington, IN; RC 2, Boston Scientific, Natick, MA; RIM, Cordis Endovascular, Warrenton, NJ) are performed. FMD typically involves the distal main renal artery and its branches, so at least two oblique selective angiograms should be obtained of each renal artery to define the lesions and their relationship to branch vessels. Anatomic variations are common, and careful evaluation of preprocedural cross-sectional imaging is helpful in calculating the optimal fluoroscopic projections. In addition, FMD is bilateral in more than 33% of patients, so meticulous evaluation of both kidneys and their intrarenal branches is requisite. On rare occasions a brachial artery approach is preferable when renal arteries originate from the aorta in a very caudal orientation.

Stenotic lesions should be crossed carefully using a soft-tipped wire, such as a 0.035-inch Bentson (Cook, AngioDynamics), after the selective catheter has engaged the renal artery ostium. Once the lesion is crossed by the

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