

# Renal Artery Fibromuscular Dysplasia Update

Diagnosis and management of this serious but underdiagnosed disease.

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**F**ibromuscular dysplasia (FMD) is a noninflammatory, nonatherosclerotic arterial disease that predominantly affects the renal and carotid arteries but has also been identified in almost every arterial bed.<sup>1,2</sup> FMD may cause arterial stenosis, occlusion, aneurysm, and/or dissection, but many patients are likely asymptomatic and may remain undiagnosed. FMD occurs most commonly in women who are between the ages of 20 and 60, but it can be present at any age and occurs in approximately 10% of men.<sup>2,3</sup> The prevalence of FMD in the general population is unknown; however, several lines of evidence suggest that FMD has a higher

prevalence than previously thought. Cragg and associates identified FMD in the renal arteries in 3.8% of 1,862 potential renal donors who underwent renal angiography.<sup>4</sup> Neymark and colleagues found on renal angiography that 6.6% (47/716) of potential renal donors had FMD.<sup>5</sup> If these studies are accurate and applied to the general population, and we were to assume an estimated prevalence of at least 5% within the United States population of women over the age of 18, then approximately 5 to 8 million women may have FMD.

Renal artery FMD occurs in about 70% of patients with FMD and is often bilateral. The most common mani-

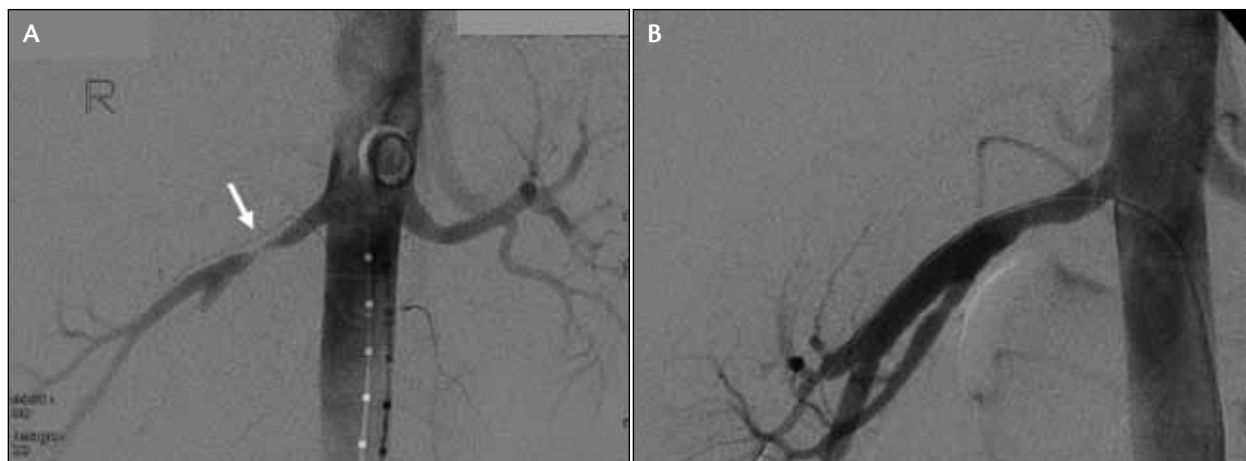
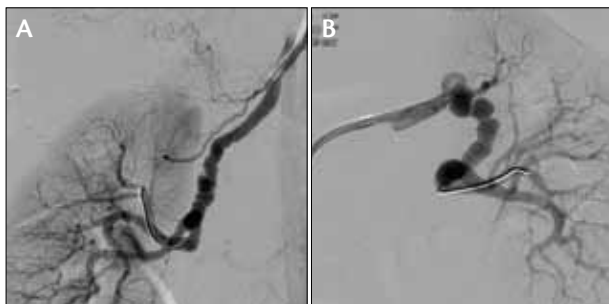


Figure 1. A 26-year-old woman with a 6-week history of severe hypertension. There is a severe focal stenosis in the mid-renal artery characteristic of intimal fibroplasia (A). Imaging after balloon angioplasty showing no significant renal artery stenosis (B). The renal artery has remained stenosis-free for 3 years, and the patient requires no antihypertensive therapy.



**Figure 2.** Medial fibroplasia of the right (A) and left (B) renal arteries. Note the string-of-beads appearance in the mid and distal renal arteries.

festation of renal artery FMD is hypertension caused by renal artery stenosis activating the renin angiotensin aldosterone axis. It has been suggested that renal artery aneurysms occur in approximately 5% to 10% of patients with FMD.<sup>5-7</sup>

### **PATHOLOGIC CLASSIFICATION OF FMD**

Regardless of which arterial bed is affected, the pathologic classification of FMD is the same. FMD is classified into three categories based on the layer of the arterial wall that is affected: intima, media, or adventitia (periarterial).<sup>8,9</sup> Medial FMD is the most common and is further divided into medial fibroplasia, perimedial fibroplasia, and medial hyperplasia.<sup>9-11</sup> There are problems with this current classification in that pathologic specimens are rarely obtained because most patients are treated with an endovascular approach. The angiographic appearance of intimal fibroplasia and medial hyperplasia are virtually identical, and adventitial disease may at times have a similar appearance. There is a need for a newer classification. Medial fibroplasia occurs in 80% to 90%<sup>2</sup> of patients with FMD. The next most common histopathologic type is intimal fibroplasia.

#### **Intima**

Histologic analysis of intimal fibroplasia reveals increased collagen deposition within the intima, with a pathologically fragmented or duplicated internal elastic lamina.<sup>9,12</sup> This leads to angiographic findings of concentric stenoses or long tubular lesions, which result from focal fibrotic band-like constrictions that often typify this FMD subtype (Figure 1).<sup>10,11</sup>

#### **Media**

Medial fibroplasia accounts for 80% to 90% of all FMD cases.<sup>2,12,13</sup> Histologic analysis reveals alternating areas of thinned media and thickened medial collagenous ridges, which create the repeating stenotic “webs” that cause arte-

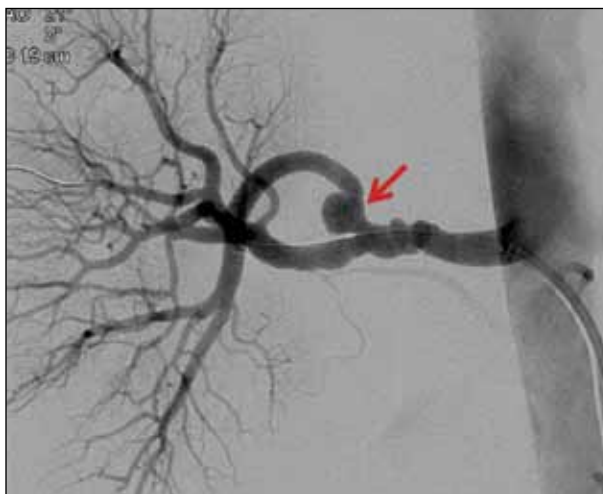
rial stenoses and poststenotic dilation that lead to the typical string-of-beads appearance on angiography (Figure 2). In a subset of patients with FMD, associated aneurysms may progressively dilate and ultimately require endovascular or surgical treatment (Figure 3).<sup>5,14</sup> Perimedial fibroplasia, which often presents with fewer and smaller “beads” than are found in medial fibroplasia, is quite uncommon and usually occurs in girls aged between 5 and 15 years who present with hypertension and renal impairment.<sup>1</sup> Medial hyperplasia is extremely rare and may be indistinguishable from intimal fibroplasias and thus requires a pathologic specimen for diagnosis.

#### **Adventitia**

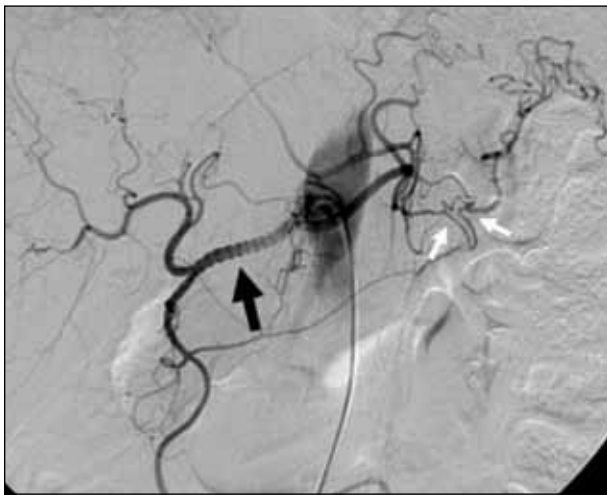
Adventitial fibroplasia has an unknown frequency of occurrence. Angiographic appearance may look similar to the intimal subtype of FMD.

### **CLINICAL PRESENTATION AND NATURAL HISTORY**

Hypertension is the most common clinical presentation in patients with FMD of the renal arteries. In young patients without obvious etiology, hypertension should trigger a search for renal artery FMD. In the FMD Registry, the mean age at diagnosis was  $51.5 \pm 14.3$  years; renal artery FMD should therefore also be considered in any patient whose blood pressure cannot be controlled on a good triple-drug regimen with one of the agents being a diuretic.<sup>7,15</sup> Earlier studies showed that the mean age for the initial diagnosis of renal artery FMD was approximately 39 in women and 31 in men. However, FMD can present at any age.<sup>16</sup> Noninvasive imaging modalities such as computed tomographic angiography



**Figure 3.** Typical medial fibroplasia and a 5.5-mm aneurysm (arrow) immediately after the bifurcation of the main renal artery.



**Figure 4. Standing or stationary waves.** Note the symmetric oscillations (black arrow) in the artery. With kind permission from Springer Science+Business Media: *Cardiovasc Intervent Radiol*, Standing waves of hepatic artery associated with renal and extrarenal fibromuscular dysplasia, 31, 2008, S38–S40, Peynircioglu B, Cil BE.

(CTA) and magnetic resonance angiography (MRA) have led to more frequent incidental findings of FMD.<sup>17</sup> Because FMD is asymptomatic in a significant number of patients, it is increasingly first identified in older patients who may also have concomitant atherosclerosis or are being imaged for another reason.<sup>3</sup> Patients may have both atherosclerotic plaques affecting the ostial and/or proximal renal arterial segments and FMD that typically manifests in the mid-distal arterial segments. Patients who are diagnosed with FMD at a more advanced age usually have had a longer duration of hypertension, and angioplasty is less likely to result in a cure of hypertension.<sup>18</sup>

Other clinical manifestations of renal artery FMD may include aneurysm, dissection, or, infrequently, occlusion of the renal artery. Dissection is the most common cause of renal infarction in patients with FMD. Although two groups have previously reported varying rates of disease progression in patients with renal artery FMD by following lesions via angiography,<sup>19,20</sup> there are still no convincing data because both of these studies had significant methodologic flaws. It remains technically difficult to assess whether a given lesion becomes more stenotic because the severity of stenosis cannot be precisely gauged by angiography or any other imaging modality except perhaps intravascular ultrasound (IVUS).

Patients with FMD may be normotensive, have mild, easily controlled hypertension, or severe and difficult-to-control hypertension. Renal dysfunction with medial fibro-

plasia rarely occurs except in the presence of a dissection and renal infarction. Intimal and perimedial fibroplasia may be associated with renal dysfunction, dissection, and progression to occlusion.<sup>2</sup>

It should be noted that approximately 65% of patients with renal artery FMD have associated carotid or vertebral artery FMD when imaging of those vascular territories is performed.<sup>7</sup> Therefore, it has been our practice to perform duplex ultrasonography (DUS) of the carotid arteries (making certain that the mid and distal carotid arteries are imaged well) and MRA of the head to exclude intracranial aneurysms in all patients with renal artery FMD.

### DIAGNOSTIC EVALUATION

Catheter-based angiography remains the most accurate imaging technique to diagnose and evaluate FMD. It can visualize the main renal arteries as well as the smaller branch vessels. Aneurysm formation and dissections in the branch renal arterial segments are also accurately evaluated with catheter-based angiography.<sup>21</sup> Furthermore, a pressure wire and IVUS imaging can be used during the procedure to help determine the hemodynamic significance of a lesion.<sup>22–24</sup> Gowda and colleagues demonstrated in a small series of patients that IVUS imaging could provide useful additive information on the intravascular morphologic appearance of FMD lesions that correlated well with routine angiographic images.<sup>24</sup> Although catheter-based angiography has been considered the gold standard for the diagnosis of FMD, the endovascular specialist should be mindful to not simply rely on angiography because stenoses related to FMD can be subtle and only detectable with pressure gradient measurements or the use of IVUS imaging.<sup>22,23</sup>

CTA and MRA have been successfully used in the diagnosis of renal artery FMD. However, neither imaging modality can accurately resolve branch vessel disease.<sup>21</sup> There are limitations to MRA in that the spatial resolution is less than CTA, and MR artifact at times can suggest the presence of beading when none really exists.<sup>2</sup> Both techniques can accurately identify aneurysms.<sup>17,25</sup>

DUS is accurate in the diagnosis of atherosclerotic renal artery stenosis and is valuable as a surveillance modality after endovascular intervention.<sup>26</sup> Although no large-scale studies have directly compared DUS with angiography, smaller reports have demonstrated the diagnostic accuracy of DUS in patients with FMD.<sup>27,28</sup> Even though the string-of-beads appearance may be seen on DUS, findings such as turbulence, vessel tortuosity, and velocity shifts within the mid-to-distal arterial segments are more commonly found. The ultrasonographer often needs to use anterior, oblique, subcostal, and flank approaches to visualize the middle and distal renal arteries.

## DIFFERENTIAL DIAGNOSIS

### Atherosclerosis

Patients with FMD can also often be distinguished from those with atherosclerotic disease due to the younger age at presentation and the lack of traditional atherosclerotic risk factors. Furthermore, atherosclerosis occurs at the ostium or proximal portion of the renal arteries, whereas FMD (particularly the most common medial fibroplasia type) occurs in the middle or distal portion of these arteries. As previously noted, it is becoming increasingly common to find older patients with both atherosclerosis and FMD.<sup>3,29-31</sup>

### Vasculitis

FMD is a noninflammatory process, whereas vasculitis involves marked inflammation of the blood vessel wall. Acute-phase reactants, such as erythrocyte sedimentation rate and C-reactive protein, are often elevated in vasculitis but usually within normal reference ranges in FMD unless there is concomitant kidney or bowel infarction. Similar to FMD, vasculitis can occur in multiple vascular territories and contribute to accelerated hypertension, kidney impairment, transient ischemic attack, or stroke. Advanced cases of vasculitis can also result in arterial stenoses and aneurysms. Although it may be difficult to distinguish multiorgan intimal fibroplasia from vasculitis, there should be no confusion between vasculitis and medial fibroplasia, as the string-of-beads appearance is distinct.

### Standing Arterial Waves

Standing arterial waves, also known as *stationary waves*, at times may be confused with FMD. They have been described as multiple serrated indentations, symmetrically distributed at evenly spaced intervals or as symmetric oscillations of the artery. Most investigators believe this is due to catheter-induced spasm (Figure 4).<sup>32,33</sup>

### Segmental Arterial Mediolysis

Segmental arterial mediolysis (SAM) is a poorly understood condition that is characterized by spontaneous dissection, occlusion, and/or aneurysm formation. Often, it is difficult to differentiate FMD from SAM. It is unclear whether SAM is a distinct vascular abnormality or a variant of FMD.<sup>34-36</sup>

Histopathologic findings include mediolysis, which begins in the outer media as a result of vacuolization. The medial arterial layer separates from the adventitial layer, increasing the likelihood of dissection. Also, arterial gaps form because of cyclical destruction and subsequent repair of the intimal and medial layer, which can result in an appearance resembling FMD. Unlike

vasculitis, there is no inflammation or fibrinoid necrosis.<sup>34</sup>

Patients with SAM may typically present with severe abdominal or flank pain caused by infarction of the visceral organs. Slavin and colleagues performed a review of 24 cases and found that of the visceral organs, the celiac artery and its branches were affected in 50% of cases. SAM affected the superior mesenteric artery or branches in 29% of cases, the inferior mesenteric artery or branches in 9%, and the renal arteries in 12%.<sup>34</sup> Men and women were equally affected, and SAM typically presents in patients between the ages of 40 and 80.<sup>35,37</sup>

## MANAGEMENT OF RENAL ARTERY FMD

The primary goal in treating patients with renal artery FMD is the control of blood pressure to prevent the sequelae of long-standing, poorly controlled hypertension.<sup>15</sup> In patients in whom high blood pressure is newly diagnosed and secondary to renal artery FMD, the initial treatment may be percutaneous balloon angioplasty (PTA).<sup>2,38</sup> The chance of curing patients so that they no longer need to take antihypertensive medications is highest when the patient is young and the duration of hypertension is short.<sup>18,39</sup>

For patients in whom FMD was not diagnosed at an early age or in whom the hypertension has already been present for many years, antihypertensive medications should be continued to maintain optimal blood pressure control. However, balloon angioplasty of the renal arteries should be considered when: (1) hypertension becomes difficult to control despite an optimal medical regimen; (2) the patient develops intolerable side effects to increasing doses of antihypertensive medications; and/or (3) renal size or function begins to decrease.

Hypertension attributed to renal artery FMD is often successfully treated with balloon angioplasty alone. There is no need for stent implantation under most circumstances. Stenting of the mid-distal arterial segments often affected by FMD may prohibit potential target sites for surgical revascularization should that option become necessary. We advocate two indications for stenting in renal artery FMD: (1) if the pressure gradient cannot be obliterated with angioplasty alone; and (2) when a renal artery dissection arises spontaneously or is created iatrogenically during intervention. The primary role for surgical revascularization is to treat aneurysms in patients in whom endovascular therapy is not an option<sup>14,40</sup> or is unsuccessful.<sup>16</sup>

PTA has supplanted surgical revascularization as the preferred treatment of renal artery FMD.<sup>16,38</sup> Angioplasty has several advantages over open surgical revascularization: it can be performed with a high degree of techni-

**TABLE 1. RESULTS OF PERCUTANEOUS TRANSLUMINAL ANGIOPLASTY IN PATIENTS WITH RENAL ARTERY FIBROMUSCULAR DYSPLASIA AND HYPERTENSION<sup>a</sup>**

Study	Year	No. of Patients	Technical Success Rate (%)	Effect on Blood Pressure (%)			Months of Follow-up (Mean [Range])	Complication Rate (%)
				Cured	Improved	Unimproved		
Sos et al <sup>41</sup>	1983	31	87	59	34	7	16 (4–40)	6
Baert et al <sup>42</sup>	1990	22	83	58	21	21	26 (6–72)	NR
Tegtmeyer et al <sup>43</sup>	1991	66	100	39	59	2	39 (1–121)	13
Bonelli et al <sup>44</sup>	1995	105	89	22	63	15	43 (0–166)	11 (major)
Jensen et al <sup>45</sup>	1995	30	97	39	47	14	12 (NR)	3 (major)
Davidson et al <sup>46</sup>	1996	23	100	52	22	26	NR	12 (minor)
Klow et al <sup>47</sup>	1998	49	98	26	44	30	9 (1–96)	0
Birrer et al <sup>48</sup>	2002	27	100	74 <sup>b</sup>		26	10 (NR)	7.4
Surowiec et al <sup>49</sup>	2003	14	95	79 <sup>b</sup>		21	NR	28.5
de Fraissinette et al <sup>50</sup>	2003	70	94	14	74	12	39 (1–204)	11
Kim et al <sup>51</sup>	2008	15	79	13	80	7	24 (1–60)	16
Davies et al <sup>38</sup>	2008	29	100	72 <sup>b</sup>		18	24 (median)	8 (minor)

<sup>a</sup>Modified and updated from Slovut DP, Olin JW. *N Engl J Med.* 2004;350:1862–1871.<sup>1</sup>

<sup>b</sup>The percentage shown is the total for cured and improved.

Abbreviations: NR, not reported.

cal and clinical success with minimal complications, it is less invasive, has a markedly shorter recovery time, is less expensive, and the procedure is now frequently performed electively on an outpatient basis. In most patients, PTA results in blood pressure reduction and can be used to treat lesions involving the main or branch arterial segments (Table 1).<sup>38,41–53</sup> Trinquart and colleagues performed a systematic review of 47 angioplasty studies (totaling 1,616 patients) and 23 surgery studies (1,014 patients) to assess the hypertension cure rate.<sup>18</sup> Cure of hypertension, defined according to the criteria in each study, was estimated to be 46% (95% confidence interval, 40%–52%) after angioplasty and 58% after surgery (95% confidence interval, 53%–62%). There was considerable heterogeneity across studies.

Most instances of recurrence of disease after PTA are related to inadequate angioplasty the first time. Usually, a second PTA results in cure or improvement in blood pressure.<sup>38,51</sup> It is important to measure pressure gradients before and after angioplasty to help assess whether all of the arterial webs have been disrupted and the pressure

gradient has been obliterated.<sup>22,23,54</sup> IVUS may be a useful adjunct to demonstrate not only the type of FMD involved but also the disruption of the webs after the intervention.<sup>22,24</sup>

Our practice is to perform a baseline duplex ultrasound within 1 to 2 weeks after angioplasty. Patients are then put into a surveillance program of ultrasound imaging performed every 6 months to assess kidney size, velocity elevations in the region of previous stenosis, and potential changes in cortical thickness. If restenosis develops without resultant hypertension, the patient is monitored serially without reintervention. Should hypertension recur and there is an area of stenosis remaining within the renal artery, angioplasty can be repeated. All patients with renal artery FMD are empirically prescribed low-dose daily aspirin therapy (81 mg).

## CONCLUSION

Although there have been significant strides made during the past several years toward educating physicians and patients about the disease, FMD still remains



frequently underdiagnosed, misdiagnosed, and inappropriately treated. Noninvasive imaging to screen for renal artery FMD should be considered if one or more of the following is found:

- Evidence for FMD in any other arterial bed, such as the carotid or vertebral arteries;
- The presence of an abdominal bruit and hypertension;
- Dissection or aneurysm of a renal, mesenteric, carotid, vertebral, or intracranial artery;
- Onset of hypertension in young individuals aged 35 or younger;
- Inability to control the blood pressure with a good triple-drug regimen. ■

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