

Diagnostic and Follow-Up Imaging for Fibromuscular Dysplasia: A Report from the United States Fibromuscular Dysplasia Patient Registry

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ABSTRACT

Background: Fibromuscular dysplasia (FMD) is a disease of medium-sized arteries that leads to beading, stenosis, dissection or aneurysm. The use of imaging for initial diagnosis and subsequent follow-up of patients with FMD has not been well-described.

Methods: Imaging data for initial diagnosis based on study date, further evaluation, and subsequent follow-up is reported for the 340 patients (pts) with renovascular or cerebrovascular FMD diagnosis enrolled in the FMD registry from 7 U.S. sites.

Results: Among pts with renal artery involvement alone. ultrasound was the most common initial diagnostic modality used (77% of patients underwent ultrasound), followed by catheter-based angiography (66.7%), CT angiography (39.7%), MR angiography (21.4%), and intravascular ultrasound (5.6%). Among pts with extracranial or vertebral involvement alone, ultrasound remained the most commonly used imaging modality (79.2%); however, fewer pts underwent catheter-based angiography (37.5%). Patients with both renal and extracranial/vertebral involvement underwent more imaging overall, with ultrasound (83.8%) and catheter-based angiography (64.1%) being the most common modalities. At follow-up, ultrasound remained the most commonly used modality among pts with renal involvement alone (72.3%), extracranial/vertebral alone (86.2%), or both (77.1%). Catheter-based angiography was also commonly used for renal alone (38.3%), extracranial/ vertebral alone (20.7%), or both (40%). 74.5% of patients who had renal FMD underwent extracranial/vertebral artery imaging, and 83.7% of patients who had extracranial/ vertebral FMD underwent renal artery imaging. Of patients diagnosed with extracranial FMD, 63.7% also underwent imaging of the intracranial vasculature.

Conclusions: Multiple imaging modalities are used to diagnose FMD, but ultrasound remains the most commonly used modality at both the time of initial diagnosis and at subsequent follow-up. The majority of pts with renal artery FMD also had imaging of the extracranial /vertebral arteries, and vice versa. However, approximately 1/3 of patients with extracranial FMD did not undergo an imaging study of the intracranial circulation to rule out intracerebral aneurysms. Further study is required to determine the optimal diagnostic and follow-up approach in this disorder.

BACKGROUND Fibromuscular dysplasia (FMD) is a non-

- inflammatory disease of medium-sized arteries that can affect any arterial bed.
- Renal and carotid arteries are the most frequently involved vessels.
- Disease can lead to beading, stenosis, dissection or aneurysm.
- Diagnosis relies heavily on the use of imaging of the affected vasculature.
- Current practices for the selection of imaging modality have not been well-described.
- Additionally, the frequency of imaging other commonly affected vascular beds once a primary diagnosis of FMD is made has not been investigated.

OBJECTIVES

- Determine the frequency of imaging modality use at the time of initial diagnosis and at subsequent follow-up for patients with renovascular and cerebrovascular FMD.
- Examine the frequency of imaging performed on additional vascular beds once FMD is diagnosed in either the renal or carotid arteries.

METHODS

- A multi-center registry of FMD patients formed in 2008.
- Michigan Cardiovascular Outcomes Research and Reporting Program (MCORRP) serves as the coordinating center.
- A standardized data collection form including diagnostic imaging tests performed is completed for each patient both at time of enrollment and at subsequent follow-up visits.
- Summary statistics presented as total number and percentages of patients.

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		Renal	Extracranial or vertebral	Renal and extracranial/ vertebral
Ultrasound				
	Diagnosis	97 (77%)	57 (79.2%)	119 (83.8%)
	Follow-up	34 (72.3%)	25 (86.2%)	54 (77.1%)
CT angiogram				
	Diagnosis	50 (39.7%)	29 (40.3%)	76 (53.5%)
	Follow-up	7 (14.9%)	6 (20.7%)	14 (20%)
MR angiogram				
	Diagnosis	27 (21.4%)	24 (33.3%)	67 (47.2%)
	Follow-up	4 (8.5%)	7 (24.1%)	21 (30%)
Catheter-based angiogram				
	Diagnosis	84 (66.7%)	27 (37.5%)	91 (64.1%)
	Follow-up	18 (38.3%)	6 (20.7%)	28 (40%)
Intravascular ultrasound				
	Diagnosis	7 (5.6%)	1 (1.4%)	4 (2.8%)
	Follow-up	7 (14.9%)	1 (3.4%)	10 (14.3%)

RESULTS

Vascular Bed Involvement vs. Imaging Modality Used at Time of Initial Diagnosis and Follow-Up

RESULTS

Frequency of Combined Imaging of the Renal and Extracranial/Vertebral Arteries

- Of the 294 patients with renal FMD, 219 also underwent imaging of the extracranial and vertebral arteries (74.5%).
- Of the 263 patients with extracranial/vertebral FMD, 220 also underwent renal artery imaging (83.7%).

Frequency of Combined Imaging of the Extraand Intracranial Vasculature

- Of the 251 patients with extracranial FMD, 160 also underwent intracranial vascular imaging (63.7%).
- Of the 35 patients with intracranial FMD, 28 also underwent extracranial imaging (80%).

CONCLUSION

- Ultrasound is the most commonly used imaging modality at time of initial diagnosis and at subsequent follow-up.
- The majority of patients with renal artery FMD also had imaging of the extracranial /vertebral arteries, and vice versa.
- However, over one-third of patients with extracranial/vertebral FMD did not undergo imaging of the intracranial vasculature to rule out intra-cerebral aneurysm.
- Further study is required to determine the optimal diagnostic and follow-up approach to FMD.

Disclosures: None