

ABSTRACT

Background: Fibromuscular dysplasia (FMD) is non-atherosclerotic, non-inflammatory vascular disease of medium-sized arteries that may result in stenosis, dissection or aneurysm. It is unclear how FMD patients who develop an arterial aneurysm differ from those who do not.

Methods: Patients enrolled in the FMD Registry from 10 U.S. sites were categorized into two cohorts: those with known arterial aneurysms and those without.

Results: Of the 615 patients in the FMD Registry, 559 patients had sufficient data for comparison. 124 (22.2%) of these 559 FMD patients reported an arterial aneurysm. The eight most common aneurysm locations were the renal (34.5%), carotid (24.5%), celiac (14.5%), aortic (13.6%), cerebral (10.9%), mesenteric (5.5%), basilar (5.5%), and vertebral arteries (3.6%). Forty patients (36.4%) had more than one aneurysm, with a maximum of 5. FMD patients with aneurysm were not found to have a greater history of headache or hypertension. There were also no significant differences between FMD patients with aneurysm and those without when we queried for family history of hypertension, aneurysm or sudden death. Within the FMD aneurysm cohort we recorded a total of 138 procedures (72 patients), of which 33.3% were balloon angioplasties alone, 15.2% were aneurysm embolizations, 12.3% were balloon angioplasties with stenting, 9.4% were renal or mesenteric artery aneurysm surgeries, and 5.8% involved covered stents for aneurysm.

Conclusions: Approximately 1 in 5 FMD patients reported an aneurysm. Male FMD patients were more likely to develop an aneurysm than female FMD patients. FMD patients with aneurysm had earlier onset of hypertension, higher prevalence of subarachnoid hemorrhage, and were more likely to have FMD identified in multiple vascular beds, especially with mesenteric and intracranial carotid artery involvement. Given the high prevalence of arterial aneurysms in this population, physicians should consider screening FMD patients for occult aneurysms. However, further research is needed to more effectively recognize FMD patients at the greatest risk for aneurysm formation and to responsibly treat those FMD patients with known arterial aneurysm(s).

Disclosures: The FMD Registry is funded by the Fibromuscular Dysplasia Society of America (FMDSA). Jeffrey Olin and Heather Gornik are non-compensated medical advisory board members to the FMDSA. Pamela Mace is a paid employee of the FMDSA.

BACKGROUND

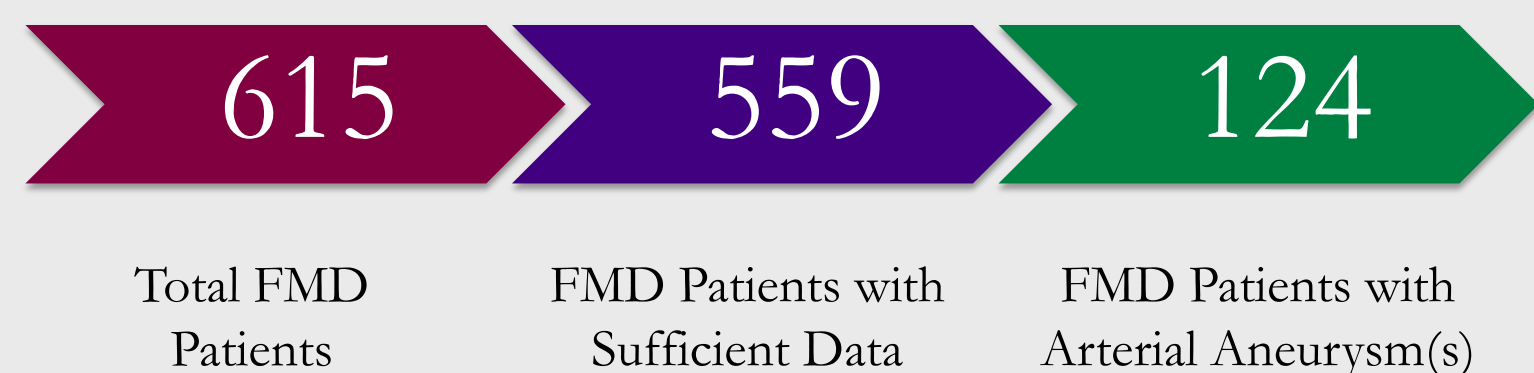
- Fibromuscular dysplasia (FMD) is an uncommon vascular disease disproportionately affecting women.
- The majority of information regarding FMD has arisen from single case reports or small case series.
- While arterial aneurysms are often regarded as complications of FMD, their incidence and details surrounding them are relatively unknown.
- Characterization of arterial aneurysms in FMD is crucial for improving the understanding, management and outcomes of all FMD patients.

OBJECTIVE

- To detail the clinical presentation, risk factors, management strategies and outcomes associated with arterial aneurysms in fibromuscular dysplasia patients.

METHODS

- The U.S. FMD Registry – formed in 2008 – consisted of 10 clinical sites and had 615 patients enrolled as of 9/11/2012.
- Michigan Cardiovascular Outcomes Research and Reporting Program (MCORRP) acts as the coordinating center.
- A standardized data collection form includes patient and family history, presenting signs and symptoms, diagnostic tests, medications, procedures and outcomes.



- 124 (22.2%) of the FMD patients with sufficient data had one or more arterial aneurysms.

RESULTS

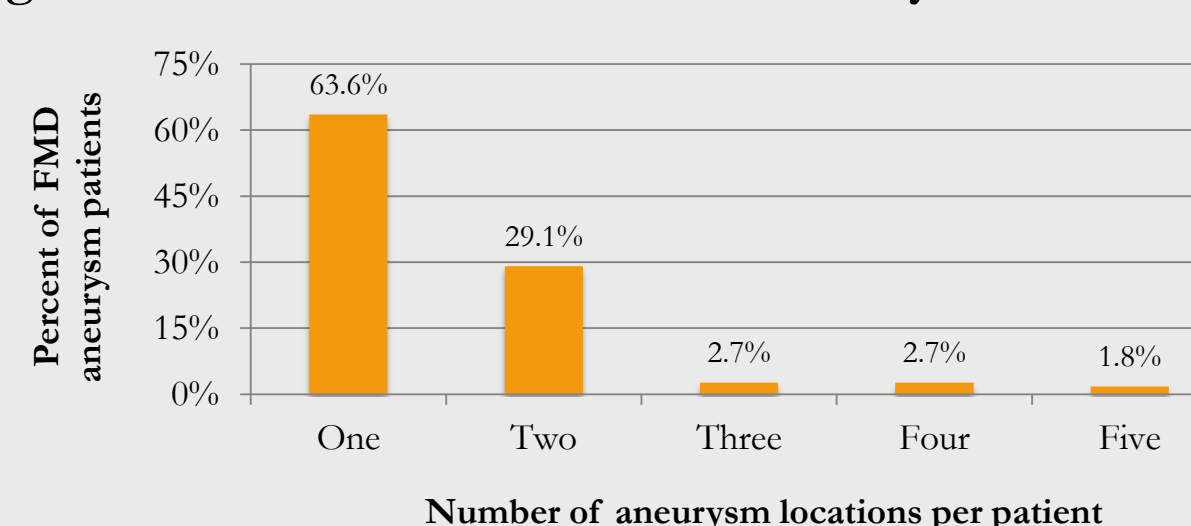
Table 1. Patient demographics, family and past medical history

	Aneurysm N=124 (22.2%)	No Aneurysm N=435 (77.8%)	p-value
Patient Demographics			
Age at diagnosis in years (Mean ± SD)	51.5 ± 15.2	52.0 ± 13.2	0.72
Male	20/124 (16.1%)	29/435 (6.7%)	0.0002
Family History			
History of hypertension	84/110 (76.4%)	295/396 (74.5%)	0.80
History of aneurysm	25/100 (25.0%)	76/383 (19.8%)	0.27
History of sudden death	14/92 (15.2%)	54/374 (14.4%)	0.87
Past Medical History			
History of hypertension	91/122 (74.6%)	304/431 (70.5%)	0.43
Age at onset of hypertension in years (Mean ± SD)	39.9 ± 16.0	44.3 ± 14.3	0.031
History of subarachnoid hemorrhage (SAH)	5/108 (4.6%)	4/419 (1.0%)	0.021
History of arterial dissection	31/111 (27.9%)	81/422 (19.2%)	0.050

Table 2. Presenting signs and symptoms

	Aneurysm	No Aneurysm	p-value
Headache	61/112 (54.5%)	231/409 (56.5%)	0.75
Hypertension	81/118 (68.6%)	265/415 (63.9%)	0.38
Cervical bruit	16/103 (15.5%)	103/387 (26.6%)	0.020
Amaurosis fugax	1/102 (1.0%)	25/405 (6.2%)	0.041
Horner's syndrome	1/103 (1.0%)	25/385 (6.5%)	0.025

Figure 1. Distribution of number of aneurysm locations (N=110)



RESULTS

Figure 2. FMD aneurysms by location (N=110)
displayed as percent of total FMD aneurysm patients with aneurysm in given location

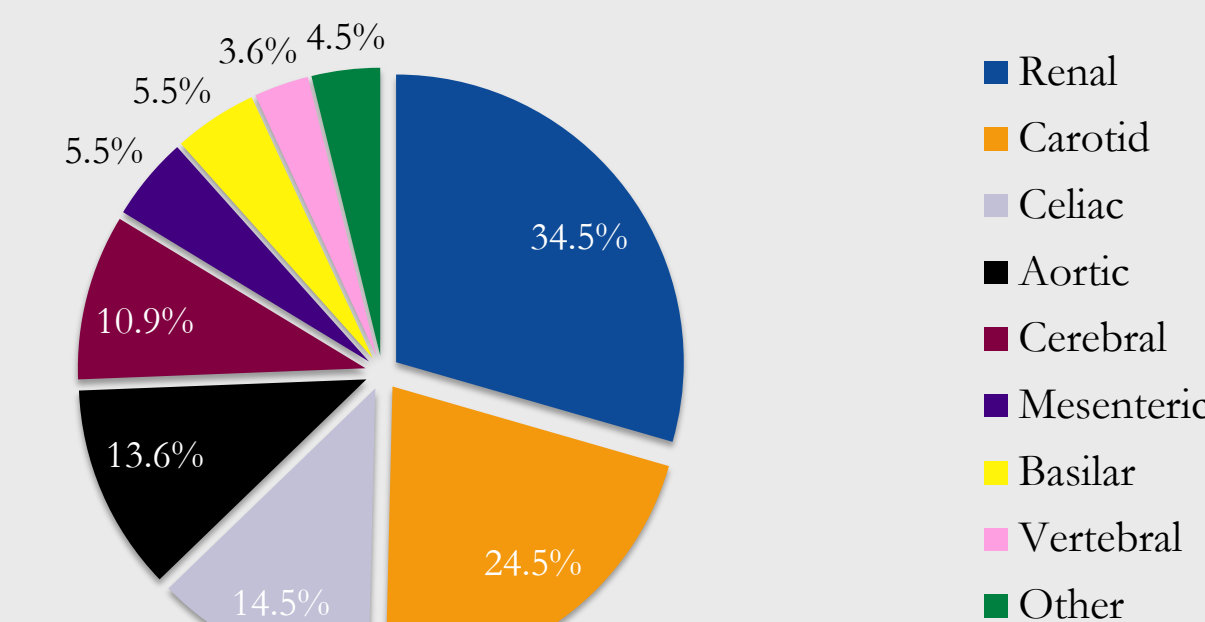


Table 3. Arterial beds with FMD involvement

	Aneurysm	No Aneurysm	p-value
Multiple arterial bed FMD	71/120 (59.2%)	190/408 (46.6%)	0.017
Extracranial carotid FMD	66/97 (68.0%)	267/354 (75.4%)	0.15
Intracranial carotid FMD	25/69 (36.2%)	18/193 (9.3%)	<0.0001
Mesenteric FMD	31/74 (41.9%)	28/194 (14.4%)	<0.0001
Vertebral FMD	25/77 (32.5%)	79/230 (34.3%)	0.78

CONCLUSIONS

- Approximately 1 in 5 FMD patients had an arterial aneurysm.
- Male FMD patients were more likely to have an aneurysm than females.
- FMD aneurysm patients had an earlier onset hypertension and were more likely to have FMD in multiple vascular beds.
- The two cohorts did not have statistically significant differences in family history or incidence of hypertension however.
- Given the high prevalence of aneurysms in this population, physicians should consider screening FMD patients for occult aneurysms.
- Still, further research is needed to better recognize FMD patients at the greatest risk of arterial aneurysm formation and to improve care for all FMD patients.