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Cleveland, Ohio

Physical Examination Findings in Fibromuscular Dysplasia: A Report from the United States Fibromuscular Dysplasia Patient Registry

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Background and Objectives

- Fibromuscular dysplasia (FMD) is an uncommon, non-inflammatory arterial disease that most frequently occurs in middle-aged women.
- Any artery can be affected, however, the renal and carotid arteries are most commonly involved.
- May lead to arterial stenosis, dissection, or aneurysm.
- Abnormal physical examination findings may be found in patients with FMD and include elevated blood pressure, arterial bruits, and neurological deficits.
- Medial fibroplasia, the most common variant, is characterized by a "string of beads" appearance on arterial imaging (Figure 1).
- Intimal fibroplasia, which is relatively uncommon, results in concentric stenosis (Figure 2).
- The objectives of this study were to determine the frequency of physical examination findings among patients with FMD as well as their diagnostic accuracy.

Figure 1: Medial fibroplasia. Arteriogram showing "string of beads" in internal carotid artery.

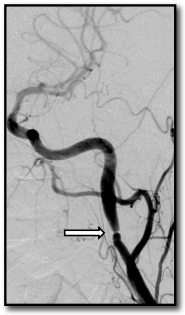

Figure 2: Intimal fibroplasia. Arteriogram showing concentric stenosis in internal carotid artery.

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 physical examination completed for each patient at the time of enrollment.
- On-line data entry environment (Drupal) into MySQL database maintained by MCORRP.
- Physical examination findings are reported for the first 447 patients enrolled from 9 U.S. clinical centers.
- FMD diagnosis confirmed with imaging for all patients (angiography, CTA, MRA, or duplex US).
- Summary statistics presented as means ± standard deviations and percentages.
- The sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) of bruits in predicting FMD were determined.

Results

Patient Demographics and Comorbidities (N = 447)	
Mean age at enrollment (years)	55.7 ±13.1
Female gender	91.0%
Caucasian	95.4%
Mean body mass index (kg/m^2)	25.5 ± 5.2
Body mass index ≥ 25 kg/m^2	48.8%
Mean blood pressure (mm Hg)	130/75 ± 20/12.4
≥ 1 Anti-hypertensive medication	77.9%
History of headache	60.0%
History of arterial dissection	18.3%
History of arterial aneurysm	17.0%

Results

Presenting Symptoms and Signs Leading to the Diagnosis of FMD (N = 447)	
Hypertension	63.8%
Headache	30.2%
Pulsatile tinnitus	27.5%
Flank/abdominal pain	15.7%
Cervical bruit	22.2%
Epigastric bruit	9.4%

Physical Examination Findings at Enrollment (N = 414 with documented physical examinations)	
Horner's syndrome (pupil abnormality or ptosis)	12.4%
Cranial nerve abnormality	9.4%
Focal neurological deficit	13.6%
Carotid bruit	30.5%
Bilateral	18.1%
Epigastric bruit	17.5%
Flank bruit	6.1%
Bilateral	3.2%
Pedal pulse deficit (dorsalis pedis and/or posterior tibial)	5.0%

Accuracy of Carotid Bruit in Predicting Extracranial Carotid FMD in Patients who Underwent Imaging

	Imaging Positive for FMD	Imaging Negative for FMD	Total
Carotid Bruit	103	5	108
No Carotid Bruit	124	74	198
Total	227	79	306

Sensitivity = 103/227 = 45.4% **Specificity = 74/79 = 93.7%**

PPV = 103/108 = 95.4% **NPV = 74/198 = 37.4%**

Results

Accuracy of Epigastric and/or Flank Bruit in Predicting Renal and/or Mesenteric FMD Among Patients who Underwent Imaging

	Imaging Positive for FMD	Imaging Negative for FMD	Total
Epigastric and/or Flank Bruit	63	5	68
No Epigastric and/or Flank Bruit	199	70	269
Total	262	75	337

Sensitivity = 63/262 = 24.0% **Specificity = 70/75 = 93.3%**

PPV = 63/68 = 92.6% **NPV = 70/269 = 26.0%**

Conclusions

- Abnormal physical examination findings are common in patients with FMD.
- Carotid bruits have a low sensitivity but high specificity in predicting the presence of cerebrovascular FMD.
- Similarly, epigastric and/or flank bruits have a low sensitivity but high specificity in predicting the presence of renal and/or mesenteric FMD.
- In the appropriate clinical setting (i.e. middle-aged women with hypertension, headaches, or pulsatile tinnitus), bruits may indicate the presence of FMD in a particular vascular bed.

Conflict of Interest Disclosure

Sponsor: Fibromuscular Dysplasia Society of America (FMDSA), a non-profit organization.

Disclosure: Drs. Gornik and Olin are volunteer medical advisory board members to the FMDSA. Ms. Pamela Mace is a paid employee of FMDSA.