



CLINICAL FEATURES AND PRESENTING SYMPTOMS AND SIGNS OF FIBROMUSCULAR DYSPLASIA: A REPORT OF THE FIBROMUSCULAR DYSPLASIA PATIENT REGISTRY

ACC Oral Contributions

Ernest N. Morial Convention Center, Room 244

Tuesday, April 05, 2011, 10:45 a.m.-11:00 a.m.

Session Title: Peripheral Arterial Disease: Update and New Therapies

Abstract Category: 11. Peripheral Arterial/Carotid Disease/Aortic Disease

Presentation Number: 925-3

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Background: Fibromuscular dysplasia (FMD), a disease of medium sized arteries, may lead to stenosis, dissection, aneurysm or occlusion. There has been little progress in understanding the epidemiology, pathogenesis and outcomes since its first description in 1938.

Methods: We report clinical features and presenting symptoms in the first 302 patients (pts) enrolled in the FMD registry from 7 U.S. sites.

Results: Most pts were female (91.4%) with a mean age at diagnosis of 51.5 + 14.3 years. The most common presenting symptoms and signs are shown below; only 4.3% were asymptomatic. The median interval from first symptom to diagnosis was within the first year (IQR 0-4 years). Family history (FH) of stroke (51.7%) or aneurysm (22.7%) was common, but FH of diagnosed FMD was uncommon (7.9%). Ultrasound was the most common diagnostic modality, followed by angiography, CTA, and MRA. The most common vascular beds involved were: renal (61.3%), extracranial carotid (55.3%), vertebral (16.6%), mesenteric (11.3%), lower extremity (8.0%), intracranial (7.3%). Medial fibroplasia was the most common type, followed by intimal disease.

Conclusions: FMD is a disorder primarily of middle-aged woman, though it presents across the lifespan and in both genders. Cerebrovascular FMD occurred at least as frequently as renal FMD. While few FMD pts had an affected family member, the prevalence of aneurysmal disease in relatives of FMD pts requires further exploration.

Common Presenting Symptoms/Signs Leading to FMD Diagnosis	
Presenting Symptom	Number Patients (%)
Hypertension	195 (64.6%)
Headache	160 (53.0%)
Dizziness	88 (29.1%)
Pulsatile tinnitus	85 (28.2%)
Cervical Bruit	74 (24.5%)
Neck Pain	65 (21.5%)
Chest Pain or Shortness of Breath	58 (19.2%)
Tinnitus	56 (18.5%)
Flank/Abdominal Pain	46 (15.2%)
Aneurysms	42 (13.9%)
Carotid Dissection	35 (11.6%)
Epigastric Bruit	31 (10.2%)
Hemispheric TIA	29 (9.6%)
Post Prandial Abdominal Pain	26 (8.6%)