› Cardiovasc Res. 2020 Apr 13;cvaa102. doi: 10.1093/cvr/cvaa102. Online ahead of print.

THE EUROPEAN/INTERNATIONAL FIBROMUSCULAR DYSPLASIA REGISTRY AND INITIATIVE (FEIRI)–CLINICAL PHENOTYPES AND THEIR PREDICTORS BASED ON A COHORT OF ONE THOUSAND PATIENTS

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PMID: 32282921 DOI: 10.1093/cvr/cvaa102

Abstract

Aims: Since December 2015, the European/International Fibromuscular Dysplasia (FMD) Registry enrolled 1022 patients from 22 countries. We present their characteristics according to disease subtype, age and gender, as well as predictors of widespread disease, aneurysms and dissections.

Methods and results: All patients diagnosed with FMD (string-of-beads or focal stenosis in at least one vascular bed) based on CTA, MRA and/or catheter-based angiography were eligible.Patients were predominantly women (82%) and Caucasians (88%). Age at diagnosis was 46±16 years (12% ≥65yo), 86% were hypertensive, 72% had multifocal and 57% multivessel FMD. Compared to patients with multifocal FMD, patients with focal FMD were younger, more often men, had less often multivessel FMD but more revascularizations. Compared to women with FMD, men were younger, had more often focal FMD and arterial dissections. Compared to younger patients with FMD, patients ≥65yo had more often multifocal FMD, lower eGFR and more atherosclerotic lesions. Independent predictors of multivessel FMD were age at FMD diagnosis, stroke, multifocal subtype, presence of aneurysm or dissection and family history of FMD. Predictors of aneurysms were multivessel and multifocal FMD. Predictors of dissections were age at FMD diagnosis, male gender, stroke and multivessel FMD.

Conclusions: The European/International FMD Registry allowed large-scale characterization of distinct profiles of patients with FMD and, more importantly, identification of a unique set of independent predictors of widespread disease, aneurysms and dissections, paving the way for targeted screening, management and follow-up of FMD.

Translational perspective: Fibromuscular dysplasia (FMD) is nowadays considered as a systemic arterial disease, warranting brain-to-pelvis vascular imaging in all patients. However, most current evidence is derived from a limited number of expert centres. Furthermore, one size may not fit all. Based on analysis of the first thousand patients enrolled in the European/International FMD registry (46 centres; 22 countries) we characterized distinct patient profiles according to FMD subtype, age and gender and identified predictors of widespread disease, aneurysms and dissections, paving the way for individualized management and follow-up. Further studies will allow refining patient characterization according to ethnicity, genetic profile and imaging biomarkers.

Keywords: Fibromuscular dysplasia; aneurysm; dissection; renovascular hypertension; stroke.