First international consensus on fibromuscular dysplasia

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The "First international consensus on the diagnosis and management of fibromuscular dysplasia" (FMD) has been published online first today in *Vascular Medicine* and the *Journal of Hypertension*. The consensus document was written by a committee of international experts in the field commissioned by the Society for Vascular Medicine (SVM) and the Working Group "Hypertension and the Kidney" of the European Society of Hypertension (ESH). The writing committee was co-chaired by Dr. Heather Gornik (Cleveland, OH, USA) of SVM and Dr. Alexandre Persu (Brussels, Belgium) of ESH and includes leading clinical experts and researchers in the field of FMD.

FMD is a nonatherosclerotic arterial disease caused by abnormal cellular proliferation and distorted architecture of the arterial wall, primarily in the renal and extracranial carotid and vertebral arteries. Clinical signs of FMD have recently been expanded to include arterial dissection, aneurysm, and tortuosity, in addition to the classical "string of beads" (multifocal) or focal arterial stenoses. Approximately 80-90% of patients with FMD are women.

This comprehensive review of FMD provides a harmonization and update to two prior European and US scientific statements and covers pathophysiology, nomenclature, differential diagnosis, diagnostic evaluation, management, and longitudinal follow-up as well as the current status of FMD registries and FMD patient advocacy organizations around the world. Physicians will appreciate the point-by-point consensus-based recommendations for managing patients with

FMD. Some highlights of the consensus points include:

- At least one focal or multifocal arterial lesion on imaging is required to establish the diagnosis of FMD. The presence of aneurysm, dissection, or tortuosity alone is inadequate to establish the diagnosis.
- Regardless of initial site of vascular bed involvement, patients with FMD should undergo brain to pelvis imaging, at least once and usually with CTA or contrast-enhanced MRA, to identify other areas of FMD, as well as to screen for occult aneurysms and dissections.
- Treatment with antiplatelet therapy (aspirin 75-100 mg/day) is reasonable to prevent thrombotic and thromboembolic complications, in the absence of contraindication.
- A standardized consensus-based protocol for renal angiography and angioplasty is proposed, which includes hemodynamic assessment of FMD lesions using translesional pressure gradient measurements.

The writing committee has also identified its top 10 research priorities for FMD. The list includes genetics, pathophysiology, epidemiology and natural history, and treatment. The list is intended to motivate basic, clinical, and translational investigators as well as scientific organizations to focus on FMD research and funding initiatives.

According to writing committee Co-Chair and SVM President, Dr. Heather Gornik, "The coming together of international experts to outline unified, consensus-based standards of clinical care for patients with FMD and an agenda to drive future research into this important but poorly understood vascular disease was our priority. We also hope that this document will provide a single go-to source for both state-of the-art and practice information on FMD for all clinicians, whether they practice in Europe, the United States, or anywhere in the world."

Dr. Persu, Co-Chair of the writing committee and Chair of the Working Group "Hypertension and Kidney" of the ESH adds: "It is an important message for patients suffering from FMD that experts from both sides of the Atlantic and elsewhere in the world stand together and speak with one voice, and that whatever the initially affected vascular bed and the specialty of the caring physician, wherever in the world, a common, comprehensive diagnostic and management strategy can be proposed. Beyond clinical practice, we also hope that this document will stimulate young researchers to engage in the field. We have now all the tools to dig further into the etiology and pathophysiology of FMD. I am convinced that such knowledge will not only help patients with FMD, but also shed light on mechanisms involved in other non-atherosclerotic arterial diseases, which jointly represent a substantial burden in young-middle aged subjects, particularly women".

More information: Gornik HL, Persu A, Adlam D, Aparicio LS, Azizi M, Boulanger M, Bruno RM, de Leeuw P, Fendrikova-Mahlay N, Froehlich J, Ganesh SK, Gray B, Jamison C, Januszewicz A, Jeunemaitre X, Kadian-Dodov D, Kim ESH, Kovacic JC, Mace P, Morganti A, Sharma A, Southerland AM, Touzé E, van der Niepen P, Wang J, Weinberg I, Wilson S, Olin JW, Plouin P-F, on behalf of the Working Group 'Hypertension and the Kidney' of the European Society of Hypertension (ESH) and the Society for Vascular Medicine (SVM)First International Consensus on the diagnosis and management of fibromuscular dysplasia. Vasc Med 2019 [Epub 16 January 2019]. DOI: 10.1177/1358863X18821816

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