

FIRST INTERNATIONAL CONSENSUS ON THE DIAGNOSIS AND MANAGEMENT OF FIBROMUSCULAR DYSPLASIA

Short title: International Consensus on Fibromuscular Dysplasia

Online Supplemental Material

Supplemental Table 1. Summary of International Consensus Key Points

- Arterial lesions of FMD should be classified according to angiographic appearance as *focal FMD* or *multifocal FMD*.
- The presence of at least one focal or multifocal arterial lesion is required to establish the diagnosis of FMD. The presence of aneurysm, dissection, or tortuosity alone is inadequate to establish the diagnosis.
- If a patient has a focal or multifocal lesion in one vascular bed to establish the diagnosis of FMD, the presence of aneurysm, dissection, or tortuosity in another/other vascular bed is considered multivessel involvement of all affected vascular beds.
- There are currently no genetic tests that are specific to FMD, and there is no justification for genetic testing of asymptomatic relatives of patients with FMD at this time. Pending future genetic developments, relatives of patients with FMD should only undergo clinical examination and imaging-based evaluation of potentially affected arterial beds upon presentation with suggestive symptoms or signs of FMD (Tables 1 and 2).
- An imaging-based evaluation for renal artery and/or cerebrovascular FMD should be considered in the presence of symptoms or signs listed in Tables 1 and 2.
- For patients with suspected renal artery FMD, CTA is the initial imaging modality of choice. Contrast-enhanced MRA is an alternative to CTA when CTA is contraindicated. Duplex ultrasound may be used as the first diagnostic procedure for renal FMD only in specialized centers with extensive expertise in duplex ultrasound for FMD.
- There are inadequate data to recommend one imaging modality over another for assessment of suspected cerebrovascular FMD. At most centers, CTA or contrast-enhanced MRA is the initial diagnostic modality of choice, as determined by local resources and experience. In high volume centers with extensive expertise in duplex ultrasound for FMD, this modality may be used for

initial assessment of suspected carotid artery FMD, though carotid duplex is not adequate to assess the distal internal carotid, vertebral, or intracranial arteries.

- Regardless of initial site(s) of vascular bed involvement, patients with FMD should undergo at least one-time assessment for intracranial aneurysm with brain CTA or MRA. Whether brain CTA or MRA should be repeated after a period of time for patients without detected aneurysm on the initial study is unknown.
- Regardless of the initial site of vascular bed involvement, patients with FMD should undergo imaging of all vessels from brain to pelvis, 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.
- In the absence of contraindication, antiplatelet therapy (i.e., aspirin 75-100 mg daily) is reasonable for patients with FMD to prevent thrombotic and thromboembolic complications.
- A consensus-based protocol for catheter-based angiography and angioplasty for renal FMD is proposed (Table 6). It is intended that this protocol will allow for standardization of diagnostic angiograms for renal FMD and for angioplasty procedures, and that prospective data will be collected to validate its use.
- Patients who have had SCAD should undergo imaging of all vessels from brain to pelvis, at least once and usually with CTA or contrast-enhanced MRA, to assess for FMD and other non-coronary arterial abnormalities.
- Patients with FMD should be seen in follow-up at least annually. Follow-up includes clinical assessment, assessment of renal function (for renal artery FMD), and imaging. At this time, there is insufficient data to recommend specific algorithms for modality and frequency of imaging studies in follow-up of FMD. The timing of follow-up imaging should be customized to each patient's pattern and severity of disease, including need for monitoring of aneurysms or dissections or following revascularization, as well as local imaging resources and experience.