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REVIEW

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# Visceral Fibromuscular Dysplasia: From asymptomatic disorder to emergency

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**Abstract**

Fibromuscular dysplasia (FMD) is an idiopathic, segmental, non-atherosclerotic and non-inflammatory disease of the musculature of arterial walls, leading to stenosis of small and medium-sized arteries, mostly involving renal and cervical arteries. As a result of better and more systematic screening, it appears that involvement of the splanchnic vascular bed is more frequent than originally assumed. We review epidemiology, pathogenesis, clinical picture as well as diagnosis and treatment of visceral artery (VA) FMD. The clinical picture is very diverse, and diagnosis is based on CT-, MR- or conventional catheter-based angiography. Involvement of VAs generally occurs among patients with multivessel FMD. Therefore, screening for VA FMD is advised especially in renal artery (RA) FMD and in case of aneurysms and/or dissections. Treatment depends on the clinical picture. However, the level of evidence is low, and much of the common practice is extrapolated from visceral atherosclerotic disease.

**KEYWORDS**

fibromuscular dysplasia, visceral

## 1 | INTRODUCTION

Fibromuscular dysplasia most commonly affects renal as well as extracranial carotid and vertebral arteries. However, it has been described in almost any arterial bed, including visceral, lower and upper extremity and intracranial arteries and even in coronary arteries. Involvement (especially symptomatic) of VAs is rare, or perhaps rather rarely reported. The disease often involves multiple vascular territories in an individual patient and may cause not only stenosis (multifocal [string-of-beads] or focal [single stenosis]), but also occlusion, dissection, aneurysm and arterial tortuosity.[1-4] FMD may be symptomatic as well as clinically silent and discovered incidentally.

## 2 | EPIDEMIOLOGY OF VISCERAL FMD

The prevalence of VA FMD in the general population is low but probably underestimated. In a review of 1100 FMD cases from the literature, only 31 presented with visceral involvement (2.8%).[5] This may be the result of underreporting or underdiagnosing as a consequence of limited knowledge of the disease and limited availability of CT scan in 1982. A literature review by Mitchell revealed only 75 well-documented cases of VA FMD in adults published since 1963.[6] In the first and in the follow-up report of the US FMD registry, including, respectively, 447 and 874 patients, mesenteric involvement was reported in, respectively, 52 patients of 198 (26.3%) and 85 patients of 528 (16.1%) who underwent imaging of this particular vascular bed.[7,8] As the entire splanchnic circulation was not imaged in all patients, prevalence may have been underestimated, but may also be confounded by indication and overestimated. In the prospective ARCADIA (Assessment of Renal and Cervical Artery Dysplasia) registry, all patients systematically underwent cross-sectional imaging (computed tomographic angiogram [CTA] or magnetic resonance angiogram [MRA]) of cervical/intracranial and abdominal vascular beds. Patients with RA FMD underwent CTA or MRA from aortic arch to intracranial vessels, and patients with extracranial cerebrovascular FMD underwent CTA or MRA from diaphragm to pelvis.[9] Involvement of mesenteric and splenic arteries was observed in 82 of 469 patients (17.5%), while in the earlier patient cohorts and the first data of the European registry, the overall prevalence of VA FMD ranged between 9% and 48.7%[3,7-15] (Figure 1).

Visceral involvement was most often reported in the mesenteric artery (MA) (14/31), followed by the coeliac trunk (10/31), hepatic- (5/31), splenic and gastric artery (each 1).[5] Systematically performed CTA to screen for aortic and/or VA abnormalities in 113 diagnosed FMD patients revealed involvement of the splenic, coeliac and superior mesenteric artery (SMA) in, respectively, 16 (14%), 15 (13%) and 10 patients (9%)[16] (Figure 2). Coeliac involvement may be overestimated as the median arcuate ligament can compress the coeliac trunk. Imaging will then

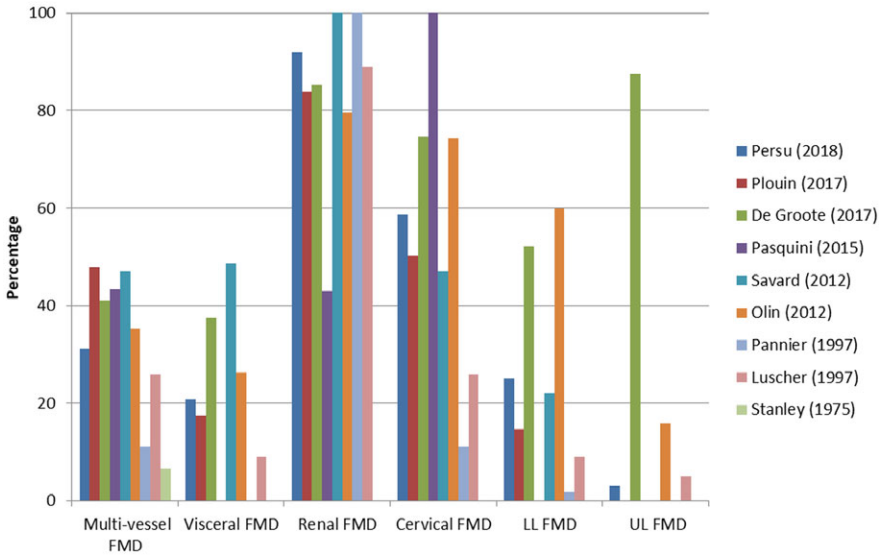


**FIGURE 1** Frequency (%) of multi-vessel, visceral and other vascular beds involvement in patients with FMD from registries and cohorts of FMD patients. The frequency percentages correspond to the ratio of patients with FMD lesions of the corresponding vascular beds to the number of patients imaged for these vascular beds. This may lead to a substantial overestimation, as radiological imaging were neither systematic nor standardised for all vascular beds, and imaging of rarely involved vascular beds was oriented according to symptoms. Consequently, the figures are biased by indication, as shown by the prevalence rate of visceral involvement of only 17.5% in the ARCADIA Registry,[9] where all patients were systematically screened using a standardised approach.[3,7,9-15] FMD, fibromuscular dysplasia; LL, lower limb; UL, upper limb

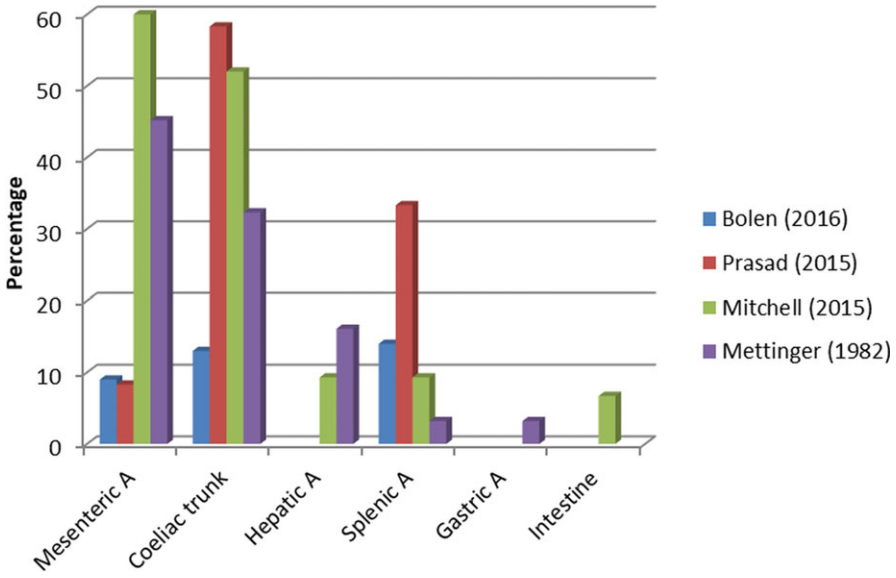


**FIGURE 2** Angiographic (and CTA/MRA) subtypes of visceral FMD involvement. The frequency percentages correspond to the ratio of patients with FMD lesions of the corresponding vascular beds to the number of patients imaged for these vascular beds. In the studies of Bolen et al, and Prasad et al, all patients were systematically screened using a standardised approach. Mitchell et al and Mettinger reported the number of published cases.[5,6,16,18] A, artery

demonstrate a focal stenosis with a characteristic hooked appearance due to indentation of the coeliac trunk on its superior surface.

Extracoronary FMD among patients with spontaneous coronary artery dissection (SCAD) has been identified through evaluation of extracoronary vessels with cross-sectional imaging from head to pelvis. Prevalence rates range from 17% to 86% depending on patient population, number of imaged vascular beds and type of imaging used for screening.[17] In the Mayo SCAD cohort, 45% of 115 patients had FMD, and 12 of 95 (12.6%) systematically screened patients had visceral involvement (7 coeliac trunk dilatation, 4 splenic artery aneurysm, 1 SMA dissection).[18]

## 3 | VASCULAR BED INVOLVEMENT AND TYPE OF FMD

In general, the pathognomonic "string-of-beads" aspect accounts for more than 80% of cases, and its histological substrate is medial FMD[7] (Figure 3Aa/Ab). In certain vascular beds, such as the mesenteric vessels, multifocal FMD appeared to be less common. Following Lüscher, the most often angiographic appearance in VA FMD is one of tubular stenosis, which correlates with intimal fibroplasia at pathological examination.[19] Aneurysmal forms of FMD may be seen as well (Figure 3B).[20]

## 4 | CLINICAL PRESENTATION: PRESENTING SYMPTOMS AND SIGNS

The US FMD registry has provided insights into the clinical presentation of FMD patients. However, registry data are limited by referral bias, as patients with symptoms or able to afford evaluation at registry centres are most likely to be included in registries. Perhaps as a reflection of this bias, only 5.6% of patients in the US FMD registry were asymptomatic.[7,8]

Most patients with VA FMD are asymptomatic because liver and intestine are relatively resistant to ischaemia, unless at least two of the major arteries are obstructed. On the one hand, there is a double hepatic blood supply (hepatic artery and portal vein), and on the other hand, a collateral circulation may develop through the inferior MA (Riolan's arcade) or between the coeliac branches and the SMA.[13] The most frequent clinical presentation of VA FMD is mesenteric ischaemia, caused by progressive stenotic lesions of the mesenteric territory and reflected as nausea, postprandial abdominal pain and weight loss. In the US FMD Registry, mesenteric ischaemia was present in 1.3% (6/447) of patients.[7] Severe forms have been reported in a few cases, eventually resulting in hemicolectomy.[6] Acute ischaemia and/or infarction is due to arterial dissection, or embolism from an aneurysm. Although mesenteric infarction seldom occurs, multi-organ failure and eventually death can occur.[21-23] One of the reasons may be the delay in diagnosis, even nowadays.[24] Ischaemic proctitis due to FMD of the superior rectal artery is exceedingly rare.[25] An epigastric bruit can be detected on physical examination, either isolated or within the classic triad indicative of occlusive or stenotic intestinal arterial disease. In children, the most common aetiology of intestinal ischaemia due to an arterial stenosis is FMD, and apparently, the MA is more frequently involved than in adults.[2,26,27]

Among patients in the US FMD Registry, dissection and aneurysm were identified in, respectively, 19.7% (n. 88) and 17.0% (n. 76) of 447 FMD patients. Coeliac trunk (2/88) and MAs (4/88) accounted for 6.8% of all

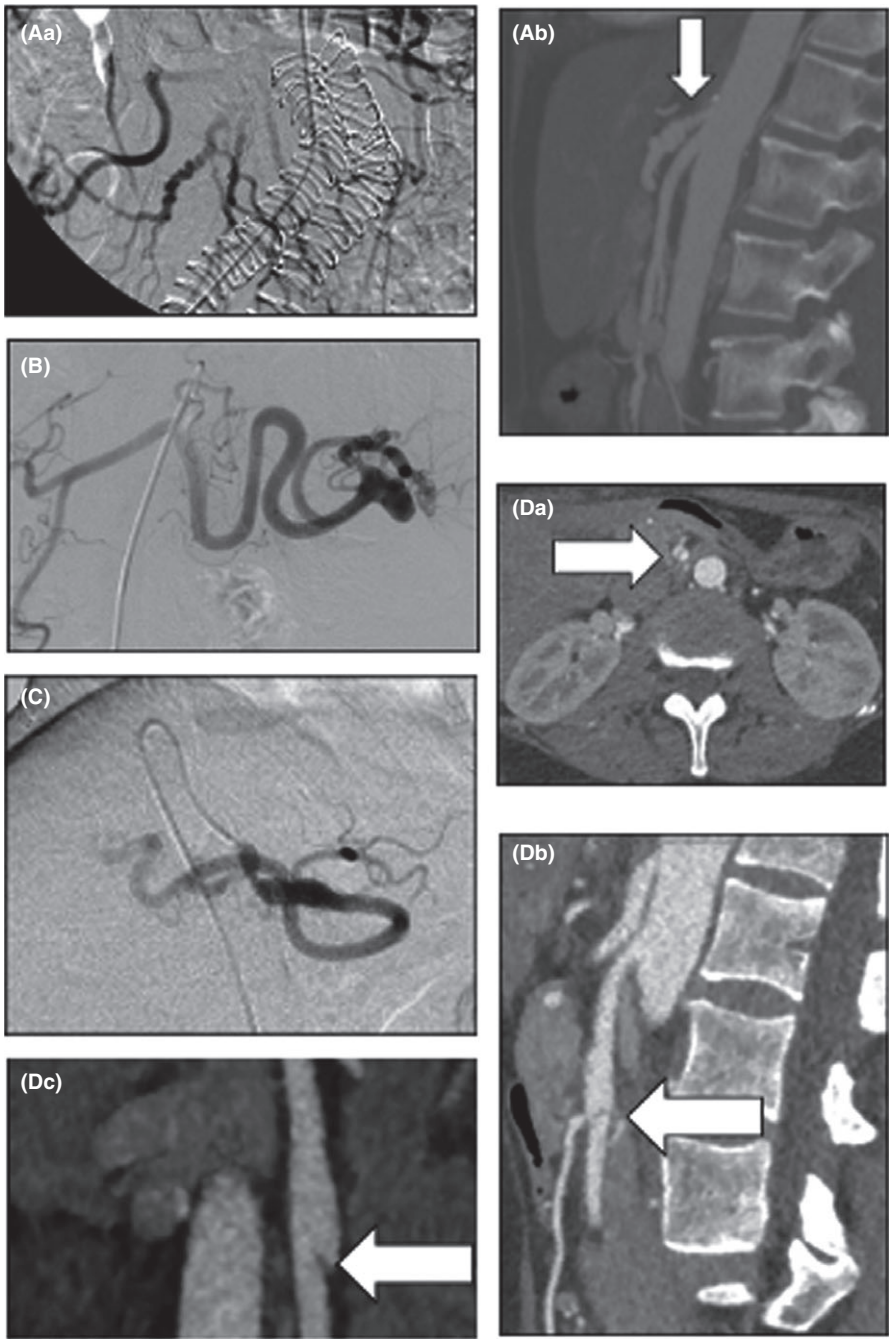


**FIGURE 3** Angiographic (catheter-based angiography and CTA) images of different FMD lesions. Panel Aa, Multifocal FMD of the superior mesenteric artery in a 82-y-old man. Panel Ab, Multifocal FMD of the coeliac trunk in a 70-y-old woman (courtesy of David Adlam, Leicester, UK). Panel B, Large aneurysm of the splenic artery in a 62-y-old woman known with multifocal FMD of right renal artery (not shown). Panel C, Stenosis of coeliac trunk and mesenteric artery (not shown) in 68-y-old woman known with multifocal FMD of both external iliac and renal arteries, and superior mesenteric artery. Panel D, Dissection of the superior mesenteric artery in a 40-y-old woman with multiple dissections and multifocal FMD of renal artery. Panel Da, axial CTA image with dissection, Panel Db: sagittal CTA image with focal dilation and intimal flap, Panel Dc: coronal CTA image with dissection. FMD, fibromuscular dysplasia; UK, United Kingdom; CTA, computed tomographic angiography

arterial dissections and 22.4% of all arterial aneurysms reported (12/76 and 5/76, respectively).[7] In a recent report of the US Registry involving 921 FMD patients, the prevalence of dissection (n. 237; 25.7%) and aneurysm (n. 200; 21.7%) has increased, likely due to the increased sample size and standardization of clinical practice and imaging. Mesenteric and coeliac aneurysms and dissections were identified in 13% and 6% of cases, respectively.[28] However, these numbers likely remain an underestimate of prevalence, as not all patients were systematically screened with imaging of all vascular beds. Splenic artery aneurysms (SAAs) are rare, but the most common (60%) of the splanchnic aneurysms[28] (Figure 3B). They occur predominantly in multiparous women.[29] Most visceral artery aneurysms (VAAs) are asymptomatic and detected incidentally on radiological studies. Physical examination can reveal an abdominal bruit and sometimes a palpable pulsatile abdominal mass. Clinical manifestations of VAAs may include abdominal pain and/or bleeding (intra-abdominal or gastrointestinal). Rupture of such aneurysms rarely occurs (<2%) but is an emergency, starting with acute and growing abdominal pain, hypovolemic shock and death. However, rupture risk in absence of pregnancy is low for aneurysms measuring <2.0 cm in diameter. Rupture of a

SAA in pregnancy is a non-obstetrical cause of abdominal pain and hemoperitoneum, resulting in high maternal and foetal mortality.[30] Multiple aneurysms can be limited to the splanchnic vascular bed, or occur in association with aneurysms in other vascular beds.[20,31]

Spontaneous MA or coeliac trunk dissections caused by FMD is rare, as reported by the US registry (respectively, 4.5% and 2.3%)[7]; however, its prevalence may have been underestimated. Several cases of visceral artery dissection (VAD) of unknown origin in patients without atherosclerotic risk factors or trauma have been reported and may be secondary to FMD. Coeliac trunk as well as SMA dissection may be asymptomatic, which may be explained by the double hepatic blood flow and the good collateral flow, but the most common presenting symptom is abdominal pain.[32,33] Subsequent thrombosis and occlusion of the coeliac trunk can result in intestinal ischaemia and hepatic failure.[34] Also in the US FMD Registry, patients with dissection (Figure 3D) were more likely to experience ischaemic events than those without dissection.[28]

## 5 | PATHOGENESIS

The pathogenesis of FMD is not known. Environmental factors have been proposed including endogenous/exogenous oestrogen exposure (oral contraceptives or hormonal replacement therapy), smoking, as well as an underlying genetic predisposition.[1,2] An association between FMD and a variant of the Phosphate and Acting Regulator 1 (*PHACTR1*) gene were identified in a case-control study involving 1154 FMD patients and 3895 controls. The presence of this gene variant increases the risk of FMD of 40% in carrier patients.[35] Recent data suggest that the pathophysiology may also be associated with abnormal regulation of TGF-beta signalling.[36] A small histological study (17 VAAs samples) using imaging mass spectrometry to assess the accumulation of lipid molecules in both the aneurysmal sac and the adjacent non-aneurysmal arteries, showed different distribution patterns of lipid molecules between FMD-associated and atherosclerotic VAAs, suggesting that diffuse accumulation of lysophosphatidylcholine, a proinflammatory and proapoptotic lipid mediator, in VAs may predispose to formation of FMD-associated VAAs.[37]

## 6 | SCREENING AND DIAGNOSIS

The optimal imaging strategy for diagnosis and surveillance of FMD lesions has still to be defined. Catheter-based angiography remains the gold standard for diagnosis, but its use is limited by its invasive character.[38] On the other hand, some FMD cases are overlooked with noninvasive imaging modalities (CTA or MRA), because mild irregularities fall below the diagnostic spatial resolution of these tests.

Screening for visceral FMD should definitely be performed in patients with symptoms suggesting ischaemia. The 2014 Expert consensus advised to consider screening of other, less often involved vascular beds in patients with renal and/or cervicocephalic FMD in the presence of suggestive symptoms or medical history.[1] The first international consensus on FMD, presented in Brussels at the International symposium "Revisiting FMD & related vascular diseases" also proposed to screen, at least once, for other areas of FMD by imaging all vessels from brain to pelvis regardless of initial site of vascular bed involvement.[39] Furthermore, in patients with FMD complications (dissection or ruptured aneurysm) in other vascular beds, there are arguments to screen for VAAs, considering the high associated morbidity and mortality.[28] Conversely, patients with VA FMD should be screened for renal and/or to a lesser degree extracranial involvement. In a survey including 75 VA FMD cases, concomitant renal or cervicocephalic involvement was found in, respectively, 41 and 3 patients. The combined involvement was more frequently present in women than in men (80% vs 36%).[6]

Diagnosis of VA FMD can be made by Duplex examination (only for chronic ischaemia), CTA or MRA, and conventional catheter-based angiography (immediately if acute ischaemia is suspected). Digital subtraction angiography, however, is able to detect even subtle vascular abnormalities, which will not be seen by CTA or MRA, despite their overall good performance. The spatial resolution of MRA is slightly inferior to CTA.[40]

Due to increasing use of endovascular treatment, tissue samples for histology are only seldom obtained. In order to attribute aneurysm or dissection to FMD, evidence of FMD lesions in another vascular bed is required. Diagnosis of FMD requires exclusion of VA spasm, median arcuate ligament syndrome (triad of postprandial abdominal pain, weight loss and often an abdominal bruit due to compression of the coeliac trunk and eventually the SMA by this ligament), trauma, atherosclerosis (but patients may have both), inflammatory arterial diseases (large artery vasculitis, ie, Takayasu arteritis, giant-cell arteritis), segmental arterial mediolysis, arterial diseases of monogenic origin (ie, vascular type of Ehlers-Danlos syndrome, neurofibromatosis type 1, …).[1,41]

## 7 | TREATMENT

To date, there are no randomized controlled studies comparing revascularization to medical treatment only, or

revascularization by PTA to surgical revascularization in FMD patients. Clinical decision making is nowadays largely guided by the evidence available in atherosclerotic disease. Usually, as recommended by the European consensus, revascularization of FMD-related lesions is considered only in cases of symptomatic FMD (eg, hypertension for RA FMD, direct or indirect signs of organ ischaemia downstream of the lesion).[1] The therapeutic decision should take into account symptomatology, type, localisation and extent of the arterial lesions, presence of associated aneurysms in the same or other territories than the primary lesion, prior vascular events related to FMD, comorbid conditions, experience of the centres, as well as age and preferences of the patient. Because of the paucity of data regarding the outcomes of endovascular and surgical interventions in FMD patients, and in the absence of evidence-based recommendations, the best therapeutic option should be discussed within a multidisciplinary team including vascular medicine experts, gastroenterologists, interventional radiologists or cardiologists trained in peripheral interventions, and vascular surgeons, all with experience of FMD management.[1,38]

For symptomatic stenosis of VAs, the first-line treatment is usually PTA, with surgical revascularization reserved for those cases where endovascular intervention is not possible.[1,38,42] For VAAs different therapeutic options are available, including conventional open or laparoscopic surgery and endovascular treatment. The choice of treatment will depend primarily on clinical presentation, aneurysm location, type of aneurysm (fusiform or saccular), associated risk factors and overall patient status.[43] Moreover, asymptomatic FMD lesions of the VAs are increasingly detected because of the frequent use of CTA (Figure 3). In general, these lesions will be conservatively managed. However, the discovery of VAAs, even if asymptomatic and irrespective of aetiology (FMD-associated or atherosclerotic), is clinically important because of the high incidence of rupture and life-threatening bleeding, with mortality rates ranging from 20% to 75% depending on the location of the aneurysm.[44] Elective intervention is recommended for all symptomatic aneurysms, for hepatic artery aneurysms (HAAs) when risk factors for rupture (multiple aneurysms and a non-atherosclerotic aetiology) are present, or for SMA aneurysms in all patients at low surgical risk because of the high rate of complications. Ligation of an aneurysm of a branch of a MA should be accompanied by resection of any ischaemic segment of bowel.[44] Ruptured aneurysms of the splenic artery require urgent action, and usually need splenectomy, as most aneurysms are located in the distal portion of the splenic artery.[45] To avoid splenectomy, ligation or transcatheter embolization can be attempted.[43,44] Elective intervention is also indicated for most aneurysms measuring >2.0 cm in diameter. Aneurysms between 1 and 2 cm in diameter should be monitored with imaging studies, initially every 6 months during the first year and if no growth is objectivated less frequent imaging can be proposed.[44] In pregnant women or women of childbearing age an aneurysm of any diameter is considered to be an absolute indication for elective repair, considering the risk of rupture with associated high foetal (95%) and maternal (75%) mortality.[42] Moreover, a literature search revealed that half of the ruptured SAAs in 32 pregnant women had a size <2 cm.[45] In the US FMD registry, about 54% and 71% of the patients with a MA aneurysm or dissection, respectively, underwent a procedure to prevent aneurysm rupture or dissection complications.[28]

There is no consensus on the optimal management strategy for VAD as well.[33,46] The goal was to prevent expansion of the false lumen leading to malperfusion and aneurysmal dilatation and rupture. If patients with VAD are asymptomatic and there are no signs of ruptured VA branches or mesenteric ischaemia, conservative treatment (anticoagulant or antiplatelet and antihypertensive therapy) may be appropriate, and follow-up with CTA or MRA is advised.[33,47-49] However, there is also no consensus on type or duration of conservative treatment. Moreover, a conservative approach may not prevent disease progression. Yet, most patients with SMAD remain asympttomatic and show improvement or no change on FU CT.[50,51] In a recent retrospective single-centre cohort of 77 patients with dissection of the SMA and/or coeliac artery, only 5% required invasive intervention because of persistent abdominal pain or bowel ischaemia. In a mean follow-up of 21 months, no late intervention or recurrence was observed.[52] Symptomatic VAD can be treated with open or laparoscopic repair surgery (vascular bypass, resection and anastomosis or ligation) or endovascular therapy.[33,46] Since endovascular therapy (coil embolization or stenting), eventual robotic system-assisted, has some advantages (ie, faster recovery, fewer complications) compared with surgery, its use has increased.[32-34] Following a recent review of the literature the initial treatment of SMAD is conservative in the majority of symptomatic patients without accompanying intestinal ischaemia or aneurysm.[53] However, an additional treatment is significantly more frequently needed in these patients than in those endovascular treated.[53] In case of persistent abdominal pain, resistant to narcotic analgesics, arterial rupture or bowel infarction, invasive therapy is mandatory.[54]

## 7.1 | Lifestyle modifications

All FMD patients should be encouraged to quit smoking[38,55,56] as smoking is the major modifiable risk factor for FMD. Savard et al showed that 30% of 337 patients with RA FMD were current smokers compared with 18% in a group of age- and sex-matched control patients with

essential hypertension ($P < 0.001$). They suggested that FMD patients, who currently smoke, have a more aggressive course with earlier-onset hypertension and subsequent increased and earlier diagnosis of FMD and are more likely to undergo renal interventions (57% vs 21% in controls).[11] Whether this association also holds true for visceral FMD is not known. Although the impact of smoking cessation on FMD progression has not been studied as such, it is a proven intervention to prevent atherosclerotic events.[57]

Besides blood pressure control, patients with prior dissection or aneurysm elsewhere should avoid exercises that may increase shear stress on the vasculature, including contact sports, heavy weight lifting, scuba diving and skydiving.[58] On the other hand, regular aerobic exercise to maintain cardiovascular health is encouraged.

### 7.2 | Pharmacologic treatment

Patients with FMD generally should receive an antiplatelet agent. However, pharmacologic treatment is merely empirical.[38] In the US FMD registry, 56.8% of the patients receive aspirin alone, 4.6% receive clopidogrel alone and 72.9% receive any antiplatelet medication.[8] No specific data for visceral FMD are reported. In case of a VAD, short-term (eg, 3-6 months) anticoagulation is generally proposed, followed by long-term antiplatelet therapy, whereas others prefer antiplatelet therapy (ie, aspirin alone or in combination with clopidogrel) for the initial treatment.[59,60] A retrospective review of 116 patients with SMAD didn't show any benefit on outcome with antithrombotic therapy.[51] Current data comparing anticoagulation and antiplatelet agents after cervical artery dissection warrant future studies.[61]

Fibromuscular dysplasia patients who have undergone endovascular interventions are prescribed antiplatelet agents, in keeping with recommendations for atherosclerotic disease.[38] Current expert opinion suggests that statins should be used only to treat dyslipidemia.[38] Patients with concomitant atherosclerotic arterial disease should be treated according to the 2013 ACC/AHA Guideline on the Treatment of Blood Cholesterol to reduce Atherosclerotic Cardiovascular Risk in Adults or the 2016 European guidelines on cardiovascular disease prevention.[62,63]

## 8 | FOLLOW-UP IN THE ABSENCE OF OR AFTER REVASCULARIZATION

After diagnosis is established, surveillance imaging of affected vascular beds has to be tailored for the individual FMD patient, depending on severity, location and nature (stenosis, aneurysm, dissection) of FMD lesions.[41] Nowadays, there is no optimal monitoring protocol in case of medical treatment alone, or post revascularization in VA FMD. If no revascularization procedure has been performed, clinical FU, at least annually, and imaging studies to evaluate progression of disease are advisable. Frequency and type of imaging study used depend on vascular bed and type of involvement and worsening symptoms. However, the utility of long-term FU CT has recently been questioned.[64] After a revascularization procedure, clinical evaluation and CTA are advised; however, timing and frequency are not well established as well.[58]

## 9 | PERSPECTIVES

Current research aimed to identify genetic and environmental factors involved in the pathogenesis, progression and clinical spectrum of FMD, as well as to assess risk of disease progression, which disease subtype is more likely to progress, involvement of other "atypical" vascular beds, occurrence of complications (dissection or aneurysms) and performance of different imaging modalities. This may lead to propose an evidence-based screening and follow-up algorithm. However, to reach these objectives, more systematically and prospectively collected data are required. Therefore, all FMD cases should be registered into national and international registries such as the ongoing French and US registries, and more recently the European FMD registry.[7,9,15]

Research, focusing on the possibility of delaying the disease or preventing the development of aneurysms by inhibitors of lysophosphatidylcholine, is ongoing.[37] Also, the role of pharmacologic agents, known to decrease vascular TGF-expression such as angiotensin receptor blockers, should be clarified.[36,65]

**TABLE 1** Take-home messages of the review

| Take-home messages |
|---|
| 1. The prevalence of visceral artery FMD among FMD patients is estimated to 15%-20% (range: 9%-50%) |
| Therefore, screening for visceral FMD is advised, especially in renal FMD as well as in case of aneurysms and/or dissections in other vascular beds |
| 2. The clinical picture is very diverse, but a triad of postprandial abdominal pain, weight loss and an abdominal bruit is pathognomonic for mesenteric ischaemia |
| 3. The gold standard for diagnosis is conventional catheter-based angiography and remains the first choice in symptomatic patients |
| CT and MR-angiography are reliable screening examinations |
| 4. Treatment depends on symptomatology, type/localisation and extent of the arterial involvement, the presence of aneurysms, prior vascular events, comorbid conditions and age of the patient |

## 10 | CONCLUSIONS

The prevalence of VA FMD may be higher than previously reported, because patients are often asymptomatic. The clinical presentation has a wide spectrum from asymptomatic lesions to critical bowel ischaemia, visceral gangrene or lethal bleeding. The classical triad including postprandial abdominal pain, weight loss and abdominal bruit, is the most common clinical presentation, indicating severe stenosis with visceral ischaemia. Screening for VA FMD in patients with renal and/or cervical artery FMD, especially in cases of aneurysm or dissection may be useful to prevent complications.

There are no specific guidelines for diagnosis and treatment of VA FMD, which is at least partly explained by the absence of randomized clinical trials. Most of the evidence derives from cohorts (French and US registries), case reports and expert opinions. In 2015, a European registry has been established.[39] Currently, it includes over 600 cases from 13 countries.[15] Diagnosis is made by CTA, MRA or conventional arteriography. However, definitive diagnosis is challenging, as VA FMD can closely mimic vasculitis as well as atherosclerosis. Treatment may include optimal medical FU and/or revascularization with PTA with or without coil embolization, aneurysm clipping or reconstructive vascular surgery. In patients with VA FMD, screening for FMD in other vascular beds, especially renal and cervicocephalic, is recommended (Table 1).

## CONFLICT OF INTEREST

The authors declare no conflict of interest.

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