

FIBROMUSCULAR DYSPLASIA



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INTRODUCTION



- **Fibromuscular dysplasia (FMD)** is an idiopathic, nonatherosclerotic, and noninflammatory arterial disease.
- Affects **small- to medium-sized** arteries.
- Result in multifocal **aneurysms, stenosis, tortuosity, and dissections**.
- Affects **young women <50 years** and accounts for **10-20% cases** of renal artery stenosis. F>M-3:1.
- Etiology : Unclear. Genetic – HLA DR-w6 /environmental – smoking.

Initial Consultation

Clinical Assessment for symptoms of FMD:

- Hypertension
- Headaches
- Pulsatile tinnitus
- Cervical or abdominal bruits
- Positive family history

If warranted, imaging evaluation to diagnose FMD:

- Carotid artery duplex ultrasound
- Renal artery duplex ultrasound
- Review all previous imaging (ultrasound, MRA, CTA, catheter-based angiogram)

If confirmed FMD diagnosis on imaging, medical treatment and recommendations for all patients:

- Blood pressure control
- Headache specialist if needed
- Counseling re: anxiety
- Avoidance of high velocity neck manipulation, high gravitational force activities
- Aspirin 81 mg daily
- Clinical follow-up biannually x 2 years, then annually

If FMD Diagnosis is Confirmed:

All patients should have cross-sectional imaging (MRA or preferably CTA) from head-to-pelvis to search for aneurysm(s) and/or dissection(s).

Aneurysm

Assess location, size, and shape

Aneurysms that do not need immediate treatment are followed every 6-12 months with a cross-sectional imaging technique.

Or, refer to a surgical or endovascular specialist for consultation and treatment

Dissection

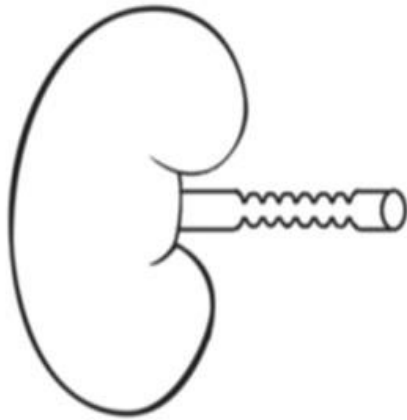
Ongoing ischemia

Refer to endovascular or surgical specialist for treatment

Absence of ischemia

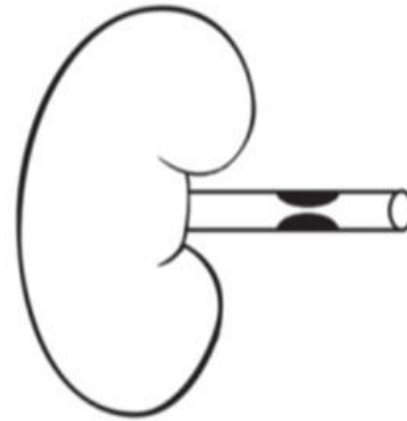
Antiplatelet or anticoagulation for 3-6 months → repeat imaging and assess symptoms.

Fibromuscular dysplasia



- Preserved renal blood flow.⁵⁸⁻⁶⁰
- Preserved glomerular filtration.^{9,51,55}
- Intact response to renin-angiotensin system modulation.⁵⁹
- Normal renin secretion in vast majority of patients.^{58,59}
- Negative correlation between renin levels and blood pressure.⁵⁸
- Revascularization cures hypertension in 40-62% of the patients.¹²

Atherosclerotic renal artery stenosis



- Reduced renal blood flow.^{58,60}
- Reduced glomerular filtration.^{57,58}
- Disturbed response to renin-angiotensin modulation.^{62,63}
- Increased renin secretion in a large subset of patients.^{58,69,70}
- Positive correlation between renin levels and blood pressure.⁵⁸
- Revascularization generally not superior to medical treatment.⁸⁶⁻⁸⁹

TREATMENT



- Anti platelet therapy- aspirin.
- Anti hypertensive medications- ACEI/ARB/CCB/B blockers.
- Statin therapy.
- Migraine therapy.
- Life style :physical activity/stress management/smoking cessation.
- Treatment based on vascular territory involvement.

TREATMENT



Renal bed

Presentations: Hypertension / dissection / renal infarcts.

Investigation: Renal angiogram / screening with CTA preferred over MRA / Duplex USG [expert].

Treatment : Revascularization for hemodynamically significant stenosis with angioplasty.

- ❖ Endovascular therapy [stenting and coiling].
- ❖ Anti platelet therapy.
- ❖ 3-6 months anticoagulation following dissection.
- ❖ Treat hypertension.
- ❖ Stop smoking.

TREATMENT



Coronary bed

Presentation :- Spontaneous coronary artery dissection[SCAD] / ACS / arrhythmias.

Investigation- CAG.

Treatment :- Conservative management in 80%.

- ❖ PCI in ongoing ischemia.
- ❖ CABG [when CAG not possible]
- ❖ Anti platelet therapy.[APT]
- ❖ DAPT [dual APT]for 1 year if stenting done.
- ❖ B blockers.
- ❖ Treat hypertension.

TREATMENT



Cerebral bed

Presentations : Pulsatile tinnitus , migraines , dizziness ,Horner syndrome , TIA.

Investigations : Screening with CTA/MRA

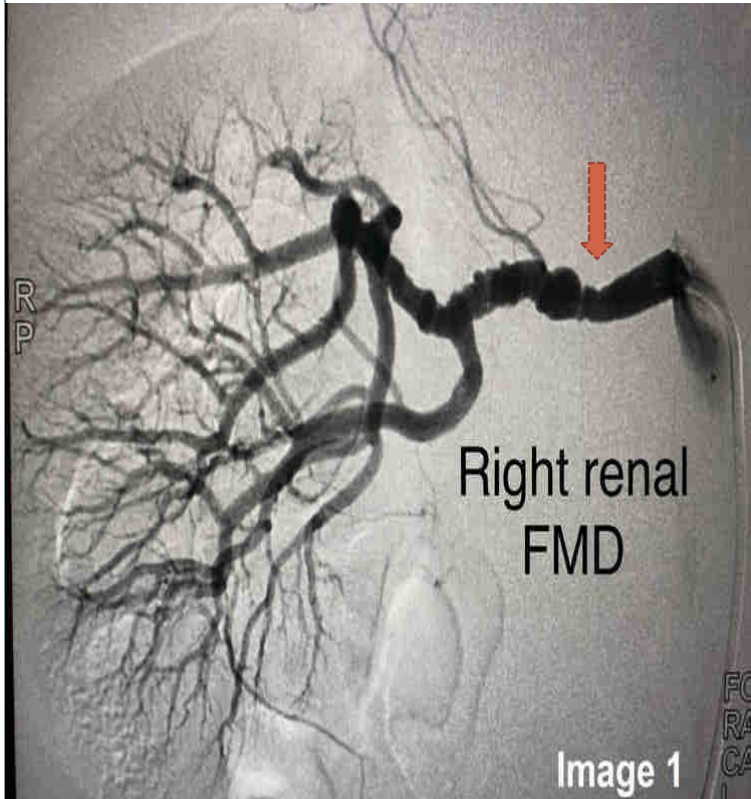
❖ Cerebral angiogram.

Treatment: Endovascular therapy.[clipping /coiling].

❖ Anti platelet therapy.

❖ Migraine treatment.

Fibromuscular Dysplasia, a Systemic Arterial Disease



String of beads appearance

Main Arterial Beds Involved

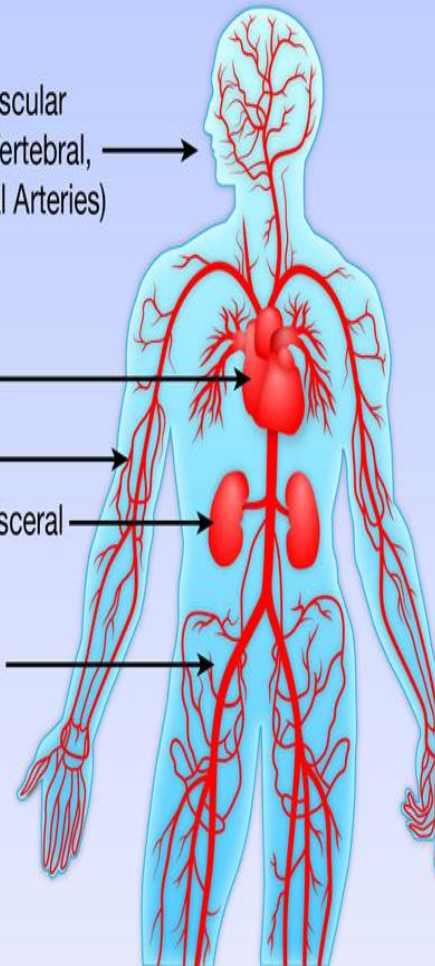
Cerebrovascular
(Carotid, Vertebral,
Intracranial Arteries)

Coronary

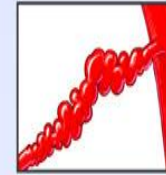
Brachial

Renal & Visceral

Iliofemoral



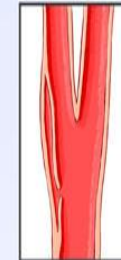
Main Arterial Manifestations



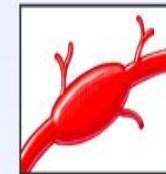
String-of-Beads*



Focal Stenosis*



Dissection



Aneurysm



Tortuosity