

Fibromuscular Dysplasia: a systemic disease

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Fibromuscular Dysplasia is a Vascular Disease distinct from atherosclerosis and inflammatory vasculitis which can involve any major artery of the body although most commonly involving renal and internal carotid arteries. Its presentation ranges from no symptoms to multi-organ involvement similar to a necrotizing vasculitis.

It involves a spectrum of multiple entities classified pathologically according to the layer of the vessel wall involved. To date involvement has only been reported in arteries where it can involve intima, media, junction between media and adventitia or adventitia as follows:

Intimal Fibroplasia: focal concentric stenoses – young male - rare

Perimedial Fibroplasia: affects junction of media and adventitia – multiple beads smaller in caliber than normal artery

Medial Fibroplasia: string of beads – young females – most common

Medial Hyperplasia: similar to intimal fibroplasias - rare

Adventitial: sharply localized tubular stenoses – very rare

Risk factors: cigarette smoking, hypertension, first degree relative, Ehlers-Danos Syndrome type IV, Alports, Pheochromocytoma, Marphan's and Takayasu Arteritis,

Male:Female 20:80

Distribution:

Renal artery	60-75%	bilateral 35%
Carotid/Vertebral:	24-30%	with intracranial aneurysm 7-50%
Visceral arteries	9%	
Peripheral arteries	5%	
>1 Vascular bed:	28%	iliac, popliteal, splanchnic, hepatic, coronary, subclavian, brachial, aorta, tibial SFA, peroneal

Imaging:

DSA remain Gold Standard

MRA Diagnostic Accuracy: older scanners 50%, newer scanners 90%

Phase contrast increase MR accuracy

Ultrasound can sometimes visualize carotid FMD

CTA accuracy is not well studied

Management: Balloon Angioplasty for stenoses Cx rate =14%, Restenosis = 7-27% at 2y

Surgery vs covered stent for aneurysms

Avoid massage or Chiropractors which can cause dissection (esp. carotids)

+/- anticoagulation

References:

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