Université de Montréal
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Winnipeg Vascular & Endovascular Symposium

Residents' Day

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Carotid Fibromuscular Dysplasia

Diagnosis, Screening & Treatment

WVES | Winnipeg Vascular & Endovascular Symposium

Presenter Disclosure

I have no current relationships with commercial entities.

WVES | Winnipeg Vascular & Endovascular Symposium

Presentation plan

1

Pre-test

2

FMD

- Generality
- Etiology
- Classification

3

Carotid FMD

- Generality
- Clinical presentation
- Diagnosis
- Screening
- Treatment

4

Post-test

Pre-test

Which of the following is considered the gold standard for diagnosis of carotid FMD ?

1. Digital substracted arteriography
2. CTA
3. MRA
4. Duplex ultrasonographic

Pre-test

Which of the following symptoms is not commonly associated with the presentation of FMD ?

1. Headaches
2. Auditory wooshing
3. Inflammatory carotidynia
4. Lightheadedness

Pre-test

What would be the appropriate treatment for an asymptomatic carotid FMD ?

1. ASA DIE
2. ASA DIE + Plavix DIE
3. ASA DIE + Angioplasty
4. ASA DIE + Angioplasty + Stent

Fibromuscular Dysplasia (FMD)

- Nonatherosclerotic, noninflammatory process
- Medium-sized vessels
- Distribution of symptomatic FMD
  - ✓ Renal artery
  - ✓ **Mid to distal internal carotid artery**
  - ✓ External iliac artery

Etiology

1. **Humoral**
   - Preponderance of **women** has been noted in all series of patients (> 90%)

2. **Ischemic**
   - Arteries involved have few branches
   - Limits the penetration of vasa vasorum

Etiology

3. <u>Mechanic</u>

- Internal carotid & renal arteries are repeatly streched during head motion & respiration
- In vitro studies demonstrates increased production of collagen, hyaluronate & condroitin sulfate du to streching

4. <u>Genetic (5%)</u>

- Increased in Caucasians, identifical twins & family members
- PHACTR1 gene

Histopathologic classification

Table 1. Pathologic and Angiographic Features of FMD

Lesion Classification	Frequency (%)	Pathologic Findings	Angiographic Findings
Intimal fibroplasia	5	Subendothelial collagen deposition in the intima with disruption of internal elastic lamina	Long, smooth, tubular stenosis in younger patient. Bilateral in majority. Progress to smooth discrete stenosis
Medial hyperplasia	1-2	Isolated smooth muscle hyperplasia without inflammation or fibrosis with normal intima and adventitia	Concentric stenosis in main renal arteries
Medial fibroplasia	80-90	Alternating areas of thickened and thinned media. Outer media with fibrous tissue. Inner media with ground substance and collagen separating smooth muscle. Intima and adventitia usually normal in all but most advanced cases	"String of beads" appearance with aneurysmal dilatation greater than normal vessel diameter alternating with web-like stenoses. Bilateral in 55%. Branch vessel disease in 25%
Perimedial fibroplasia	10-15	Accumulation of elastic tissue between adventitia and media. Normal-appearing intima	Multiple high-grade stenoses in main renal arteries without aneurysmal dilation. Progresses to occlusion

Curry TK, Messina LM. Fibromuscular dysplasia: when is intervention warranted?. *Semin Vasc Surg*. 2003;16(3):190-199. doi:10.1016/s0895-7967(03)00024-3

Classification – Radiologic findings

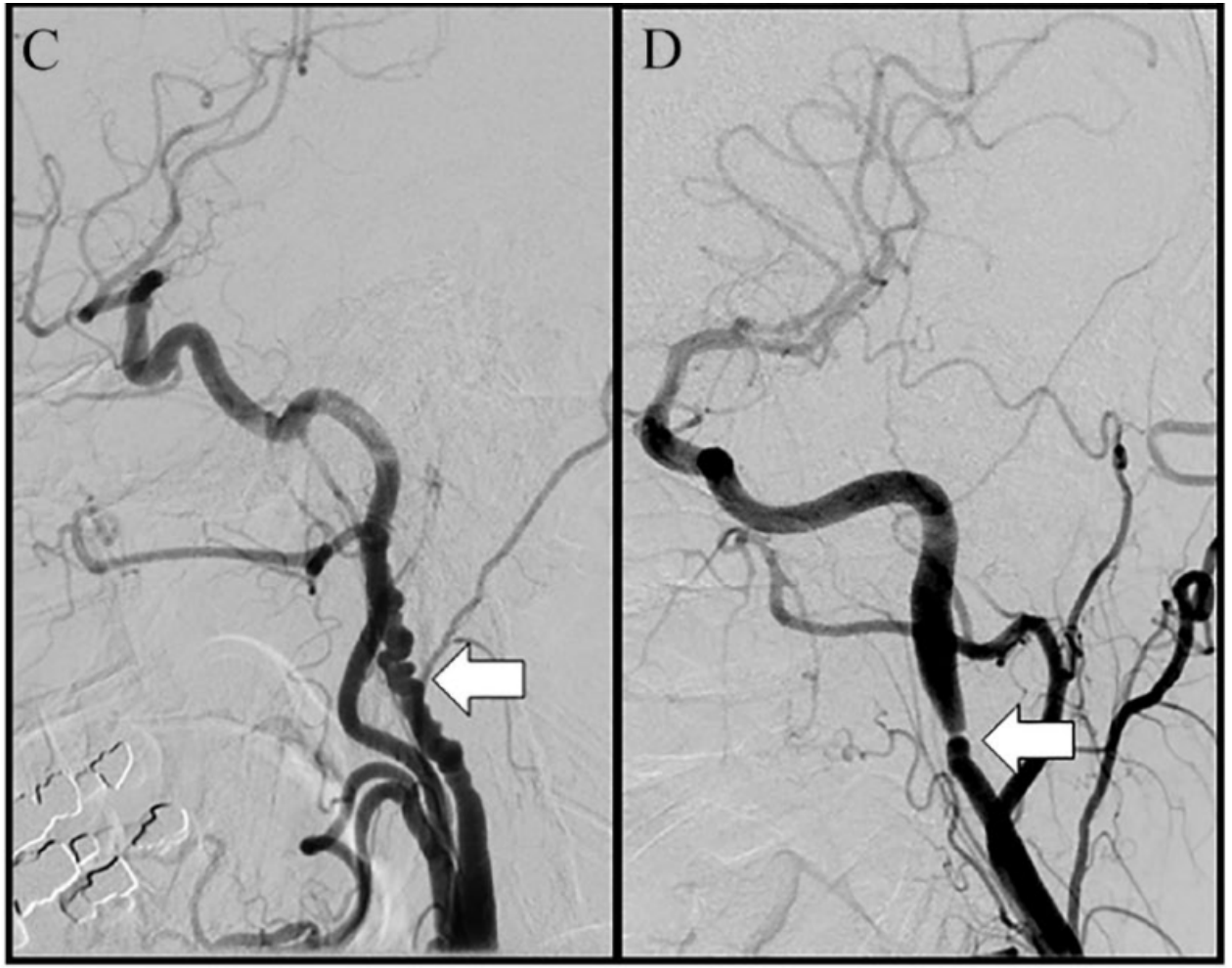
Table. Angiographic Classification of Fibromuscular Dysplasia (FMD) With 2 Categories: Focal and Multifocal[a]

Characteristic	Multifocal FMD	Focal FMD
Angiographic appearance	Alternating arterial dilatation and constriction ("string of beads" appearance); areas of dilatation are larger than the normal caliber of the artery; commonly occurs in the middle or distal portion of the artery	Unifocal concentric (<1 cm in length) or tubular (≥1 cm) smooth narrowing[b]; can occur in any portion of the artery
Histologic characteristics	Medial fibroplasia (most common) involves a functional transformation in smooth muscle cells of the arterial media, ultimately associated with plurifocal medial fibroplasia, attenuation of elastic fibers, and abnormal collagen synthesis[8]; perimedial fibroplasia (rare)[c]	Intimal fibroplasia (most common); adventitial (periarterial) fibroplasia (rare); medial hyperplasia (rare)
Frequency	90%	10% (Unifocal FMD, <10%; tubular FMD, <1%)
Associated features	Aneurysm, dissection, and tortuosity of medium-sized arteries may be present, but their presence alone is insufficient to diagnose FMD; multifocal and focal lesions may coexist in the same patient (20%)	Same as multifocal FMD

Figure 1. Multimodal Imaging of the Same Vessel of a Typical Multifocal Cervical Fibromuscular Dysplasia

Touzé E, Southerland AM, Boulanger M, et al. Fibromuscular Dysplasia and Its Neurologic Manifestations: A Systematic Review. *JAMA Neurol.* 2019;76(2):217–226. doi:10.1001/jamaneurol.2018.2848

Classification – Radiologic findings



Gornik HL, Persu A, Adlam D, et al. First International Consensus on the diagnosis and management of fibromuscular Vasc Med. 2019;24(2):164-189. doi:10.1177/1358863X18821816

FMD of the carotid artery

- First described in 1964 by Palubinskas and Ripley in *Radiology*

Fibromuscular Hyperplasia in Extrarenal Arteries[1]

A. J. PALUBINSKAS, M.D., and H. R. RIPLEY, M.D.[2]

HYPERTENSION secondary to fibromuscular hyperplasia of one or both renal arteries is now a well recognized clinical syndrome. In many instances the occlusive process is amenable to vascular reconstructive procedures.

The severity of the lesion varies. Well developed ones show prominent irregular tions of the main renal artery branches. Its typical roentgen appearance has been described in recent publications (1, 2).

Fibromuscular hyperplasia has a decided sexual affinity. Approximately 90 per cent of the cases seen at the University of California Medical Center, San Francisco, have been in females.

Prevalence

- Remains unknown
- **0.02%** of consecutive autopsies performed at the Mayo Clinic during 25 years detected carotid FMD
- 0.5% to 3% of cerebral agiograms of patients with neurological symptoms

Clinical presentation

History

- Headaches
- Severe and/or chronic migraine
- Pulsatil tinnitus (‘whoosing’ or ‘swooshing’ sound in the ears)
- Dizzines / lightheadedness
- Unilateral neck pain
- Stroke, TIA, amaurosis fugax

Physical exam

- Hypertension (renal fibromuscular dysplasia)
- Carotid bruit

Labs

- CRP
- ESR

Gornik HL, Persu A, Adlam D, et al. First International Consensus on the diagnosis and management of fibromuscular. Vasc Med. 2019;24(2):164-189. doi:10.1177/1358863X18821816

Natural History

- In a serie of 79 patients, 3 (4%) experienced a cerebral ischemic attack over a 5-year follow-up period
- In studies, one-third of lesions exhibited significant radiologic progression over time
- Currently, no established criteria exist for intervention based solely on radiologic patterns of progression

Diagnostic evaluation

VASCULAR MEDICINE

Consensus Document

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First International Consensus on the diagnosis and management of fibromuscular dysplasia

Heather L Gornik[1*], Alexandre Persu[2*], David Adlam[3,4], Lucas S Aparicio[5], Michel Azizi[6,7,8], Marion Boulanger[9], Rosa Maria Bruno[10], Peter de Leeuw[11], Natalia Fendrikova-Mahlay[12], James Froehlich[13], Santhi K Ganesh[13], Bruce H Gray[14], Cathlin Jamison[15], Andrzej Januszewicz[16], Xavier Jeunemaitre[17,18], Daniella Kadian-Dodov[19], Esther SH Kim[20], Jason C Kovacic[19], Pamela Mace[21], Alberto Morganti[22], Aditya Sharma[23], Andrew M Southerland[24], Emmanuel Touzé[9], Patricia van der Niepen[25], Jiguang Wang[26], Ido Weinberg[27], Scott Wilson[28,29], Jeffrey W Olin[19] and Pierre-Francois Plouin[6,7,8**], on behalf of the Working Group 'Hypertension and the Kidney' of the European Society of Hypertension (ESH) and the Society for Vascular Medicine (SVM)**

Diagnostic evaluation – CTA & MRA

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.

Diagnostic evaluation - Duplex

Not adequate to assess :

- Distal internal carotid
- Vertebral arteries
- Intracranial arteries

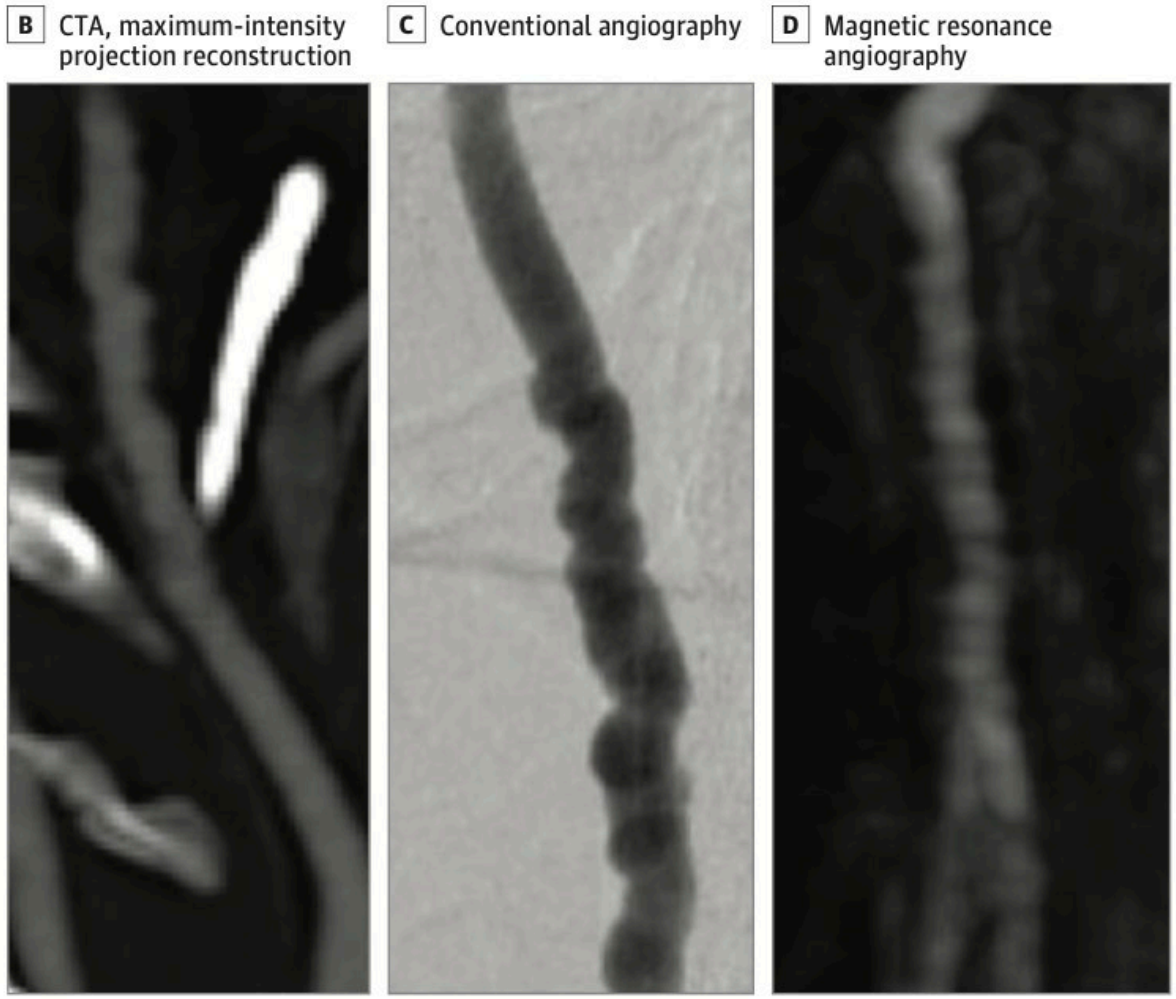
It has a restricted use in the diagnosis of fibromuscular carotid dysplasia.

Diagnostic evaluation - Arteriography

- Arteriography remains the gold standard for diagnosis in the litterature

- Restricted to severe cases of FMD requiring endovascular management because of the risk of iatrogenic dissection

Diagnostic evaluation



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Screening

Regardless of the initial site of vascular bed involvement, patients with FMD should undergo at least one-time assessment for **intracranial aneurysms** with brain CTA or MRA.

- US Registry (Lather et al., 2017) : **12.9%** of women with FMD had an intracranial aneurysm VS 3% in the general population

Screening

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.

Complications and mechanisms

Complications

- Transient ischemic attack (TIA)
- Ischemic stroke

Mechanisms

Encroachment on the arterial lumen with :

- Reduced perfusion
- Cerebral embolization of thrombus
- Dissection with secondary stenosis or aneurysmal dilatation

Treatment selection

Asymptomatic disease

- Generally benign behavior
- Medical therapy only
- Antiplatelet therapy for primary stroke prevention (ASA 80 mg DIE, Grade 2C recommendation)

Symptomatic disease

- Antiplatelet therapy
- Intervention in selected cases

Treatment selection

Treatment is appropriated in cases of

- Focal ischemic events

Durability of balloon angioplasty for carotid FDM with or without stent placement are not known.

Surgical treatment – Surgical dilatation
Historic use

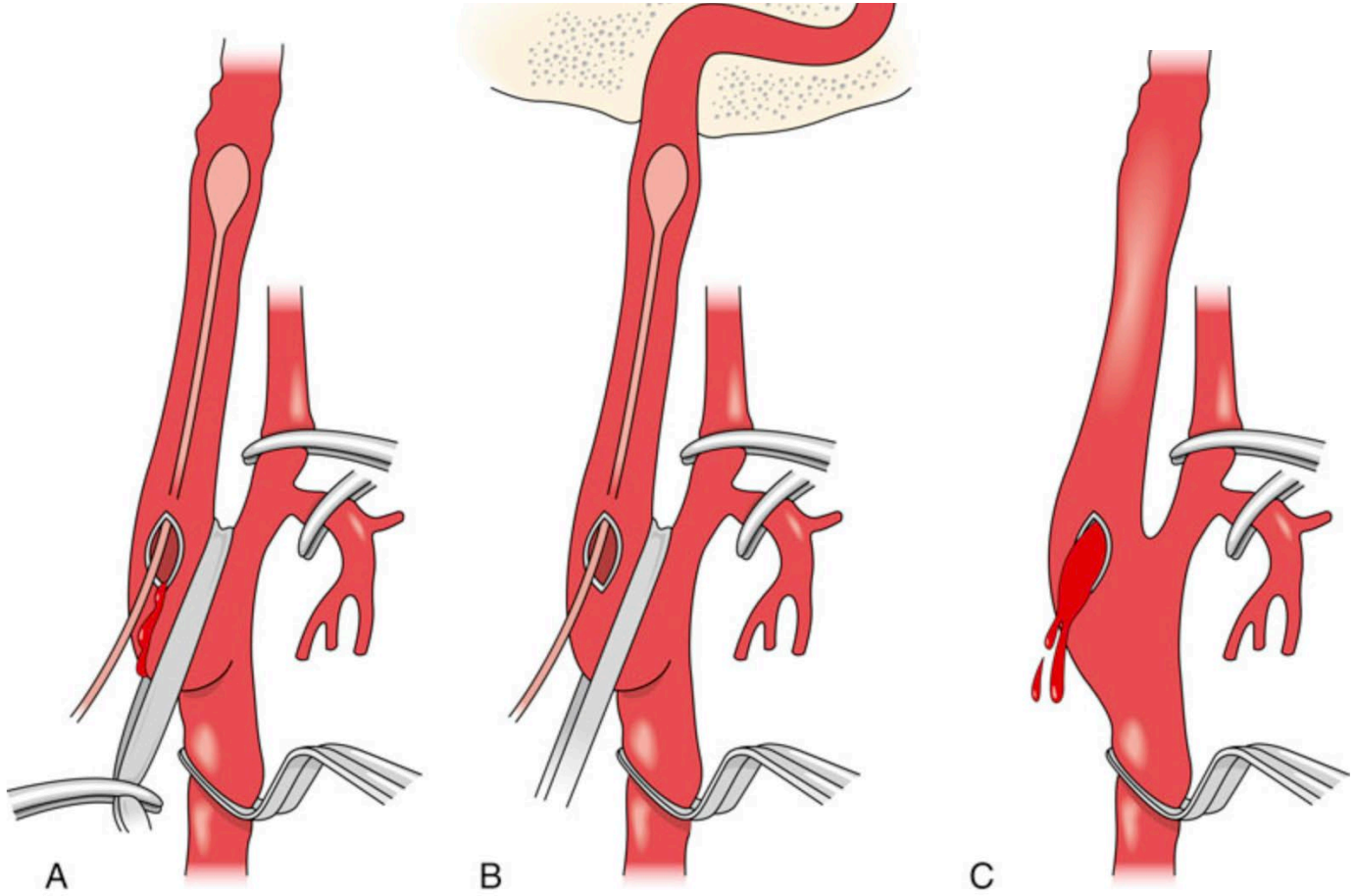

FIGURE 143.16 Drawings showing the main features of the open surgical technique. (**A**) Straightening of the carotid artery with downward traction on a polymeric silicone (Silastic) sling. (**B**) Gentle, graduated dilation of the internal carotid artery from 2 to 4 mm. Passage of the dilator to the bony canal at the base of the skull is shown. (**C**) Back-bleeding of the artery to remove dislodged debris. From Wylie EJ, et al. Nonatherosclerotic diseases of the extracranial carotid arteries. In: Egdahl R, ed. *Manual of Vascular Surgery*, New York: Springer-Verlag; 1986:184–185.

Sidawy, Anton N. & Bruce A. Perler et al. (2022). Rutherford's vascular surgery and endovascular therapy, 10th edition. Elsevier.

Surgical treatment – Balloon angioplasty

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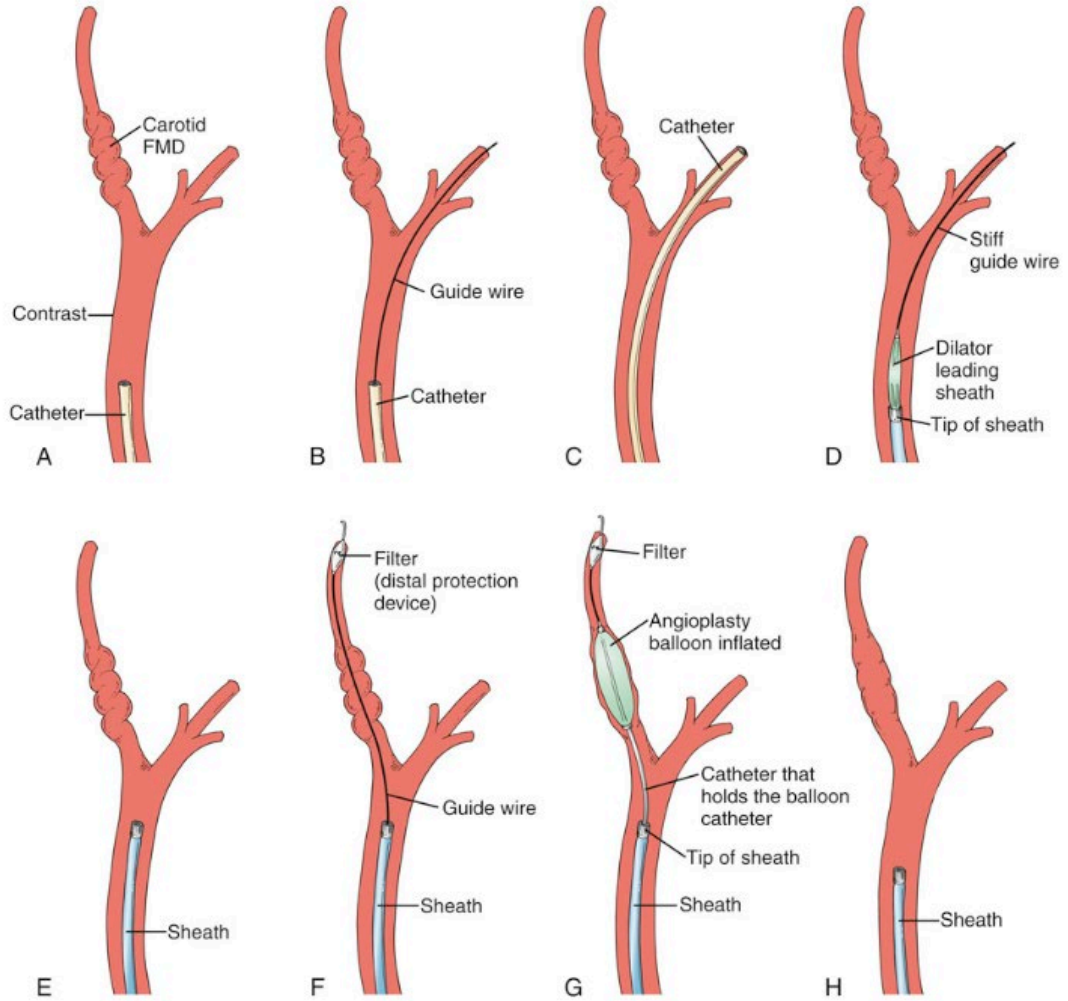

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Surgical treatment

Protection devices

- Filters
- TCAR (transcarotid artery revascularisation)

Stents

- Only if iatrogenic dissection or aneurysms
- Self-expandable stents (5 or 6 mm)

Carotid web or carotid bulb diaphragm

- Classified as atypical monofocal form of FMD of the carotid bulb
- Defined as a thin, linear, membrane that extends from the posterior aspect of the internal carotid artery bulb into the lumen
- « Intimal variant » fibromuscular dysplasia

Carotid web

VASCULAR MEDICINE

Review Article

Carotid web: Pathophysiology, diagnostic, and therapeutic options. A narrative review

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Emeraude Rivoire[1,2], Nellie Della Schiava[1,3], Olivier Rouvière[4,5], Gaele Pagnoux[4], Tae-hee Cho[6,7], Antoine Millon[1,2], and Anne Long[1,2,8]

Carotid web

Table 2. Symptomatic CaWeb: Results after open surgery or stenting.

Study type	Reference	No. of Patients	Surgery	Periprocedural complication	Clinical FU, months	Recurrent stroke or TIA	Imaging FU, months	Stent restenosis
Multicenter, retrospective observational study	Pasarikovski[59] 2024	14	CAS	Retroperitoneal hemorrhage	12[a] (IQR 10–12)	0	NA	0
Retrospective review of prospectively collected data	Brinster[4] 2023	50	CEA: $n = 27$ CAS: $n = 23$	NA NA	38[b]	0 No death	NA	NA
Retrospective	Osehobo[60] 2023	30	CAS	Hypotension $n = 19$ Bradycardia $n = 20$	17[a] (IQR 11.5–36)	0	16[a] ((IQR 9–30.5)	0
Retrospective	Marnat[57] 2023	27 (28 webs)	Dual layer CAS	Hypotension / bradycardia $n = 1$	9[a] (IQR 3.8–21.2)	0	6.1[a] (IQR 3.6–22.5)	0
Systematic review	Patel[18] 2022	151	CEA: $n = 64$ CAS: $n = 87$	NA NA	Range 3–60	0	NA NA	NA NA
Retrospective	Borghese[58] 2021	5	CEA	Local hematoma	18[b] (range 1–35)	0		
Retrospective	Haynes[61] 2020	6	CEA	Transient tongue deviation	6.1[b] ± 4	0	NA	NA
Multicenter retrospective study	Haussen[56] 2018	24	CAS	Hypotension / bradycardia $n = 2$	12[a] (IQR 3–19)	0	10[a] (IQR 3–18)	0
Systematic review	Zhang[13] 2018	70	CEA: $n = 35$ CAS: $n = 35$	NA NA	14[b] (range 6–120) 10.7[b] (range 3–144)	0 0	NA NA	NA NA

[a]Median; [b]mean.
CAS, carotid artery stenting; CEA, carotid endarterectomy; FU, follow-up; NA, not available; TIA, transient ischemic attack.

Post-test

Which of the following is considered the gold standard for diagnosis of carotid FMD ?

1. Digital substracted arteriography
2. CTA
3. MRA
4. Duplex ultrasonographic

Post-test

Which of the following is considered the gold standard for diagnosis of carotid FMD ?

1. **Digital substracted arteriography***
2. CTA
3. MRA
4. Duplex ultrasonographic

*Based on current practices, do you still consider this statement relevant?

Post-test

Which of the following symptoms is not commonly associated with the presentation of FMD ?

1. Headaches
2. Auditory wooshing
3. Inflammatory carotidynia
4. Lightheadness

Post-test

Which of the following symptoms is not commonly associated with the presentation of FMD ?

1. Headaches
2. Auditory wooshing
3. **Inflammatory carotidynia**
4. Lightheadedness

Post-test

What would be the appropriate treatment for an asymptomatic carotid FMD ?

1. ASA DIE
2. ASA DIE + Plavix DIE
3. ASA DIE + Angioplasty
4. ASA DIE + Angioplastie + Stent

Post-test

What would be the appropriate treatment for an asymptomatic carotid FMD ?

1. **ASA DIE***
2. ASA DIE + Plavix DIE
3. ASA DIE + Angioplasty
4. ASA DIE + Angioplastie + Stent

*Grade 2C recommendation

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Questions & discussion

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