Carotid-Cavernous Fistula

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CCF - dural fistula characterized by A-V shunting within cavernous sinus.

- cavernous sinus is network of venous channels traversed by intracranial portion of internal carotid artery.

ETIOLOGY

a) **head trauma** (75-80%) - ***blunt*** (esp. with temporal or sphenoid bone fractures) or ***penetrating*** (i.e. shearing or laceration of intracavernous ICA, incl. iatrogenic angiographic injury).

b) **spontaneous** (≈ 20%) - associated with [1]***ruptured intracavernous aneurysm***, [2]***fibromuscular dysplasia***, [3]***Ehlers-Danlos syndrome*** and other collagen vascular diseases, [4]atherosclerotic vascular disease, [5]pregnancy, [6]straining.

PATHOPHYSIOLOGY

- ***high-pressure arterial blood*** enters ***low-pressure venous cavernous sinus*** → interference with normal venous drainage → *compromised blood flow* within cavernous sinus (cerebral venous infarction may occur) and orbit (ophthalmic venous hypertension and orbital venous congestion).

- can be **bilateral**.

CLASSIFICATION

Direct type (70-90%):

Type A fistula - direct connection between **intracavernous ICA** and cavernous sinus.

- high-flow and high-pressure fistulas → fast progression of clinical features!!!
- more common in *young males*.
- most commonly traumatic etiology.

Dural types:

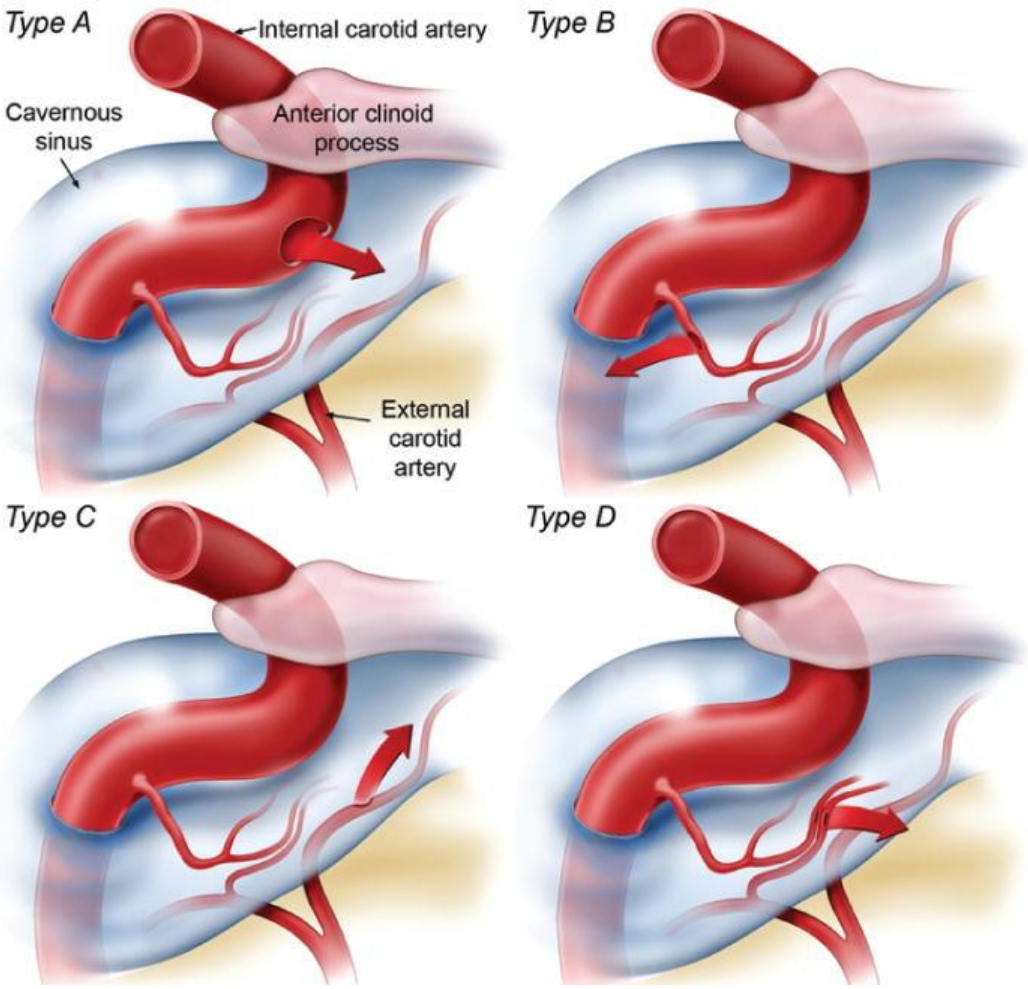
- low-flow.
- more common in *women > 50 years* (7:1 female-to-male ratio).
- most commonly spontaneous etiology.

Type B fistula - dural shunt between intracavernous branches of **ICA** and cavernous sinus.

Type C fistula - dural shunt between meningeal branches of **ECA** and cavernous sinus.

Type D fistula - **combination** of types B and C (i.e. dural shunts between ICA and ECA branches and cavernous sinus).

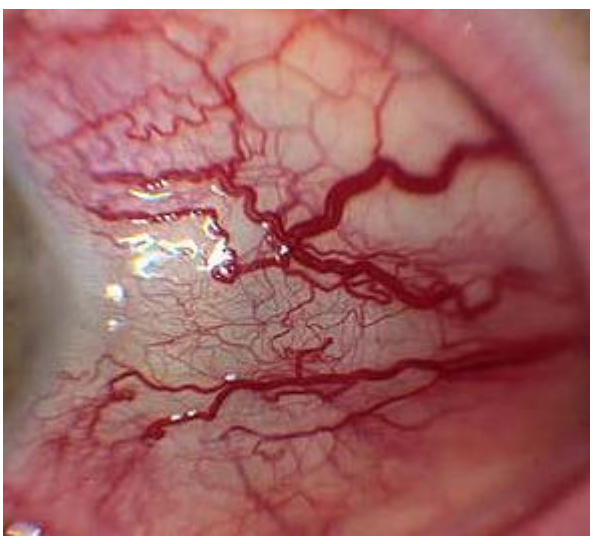
Diagrammatic representation of 4 types of fistulas:



CLINICAL FEATURES

- sudden onset:

1. Ipsilateral **ocular manifestations**:
    1) progressive pulsatile proptosis (→ corneal exposure → dehydration, traumatization), chemosis (dilated and tortuous episcleral and conjunctival vessels), arterialization of episcleral veins, edema of conjunctiva and periorbital soft tissues.
    2) cranial nerve palsy (III-VI) ipsilaterally or bilaterally.
    3) progressive (over days or weeks) monocular visual loss in late stages
    4) dilatation of retinal veins, optic disc swelling, retinopathy.
    5) central retinal vein occlusion → secondary open-angle glaucoma.



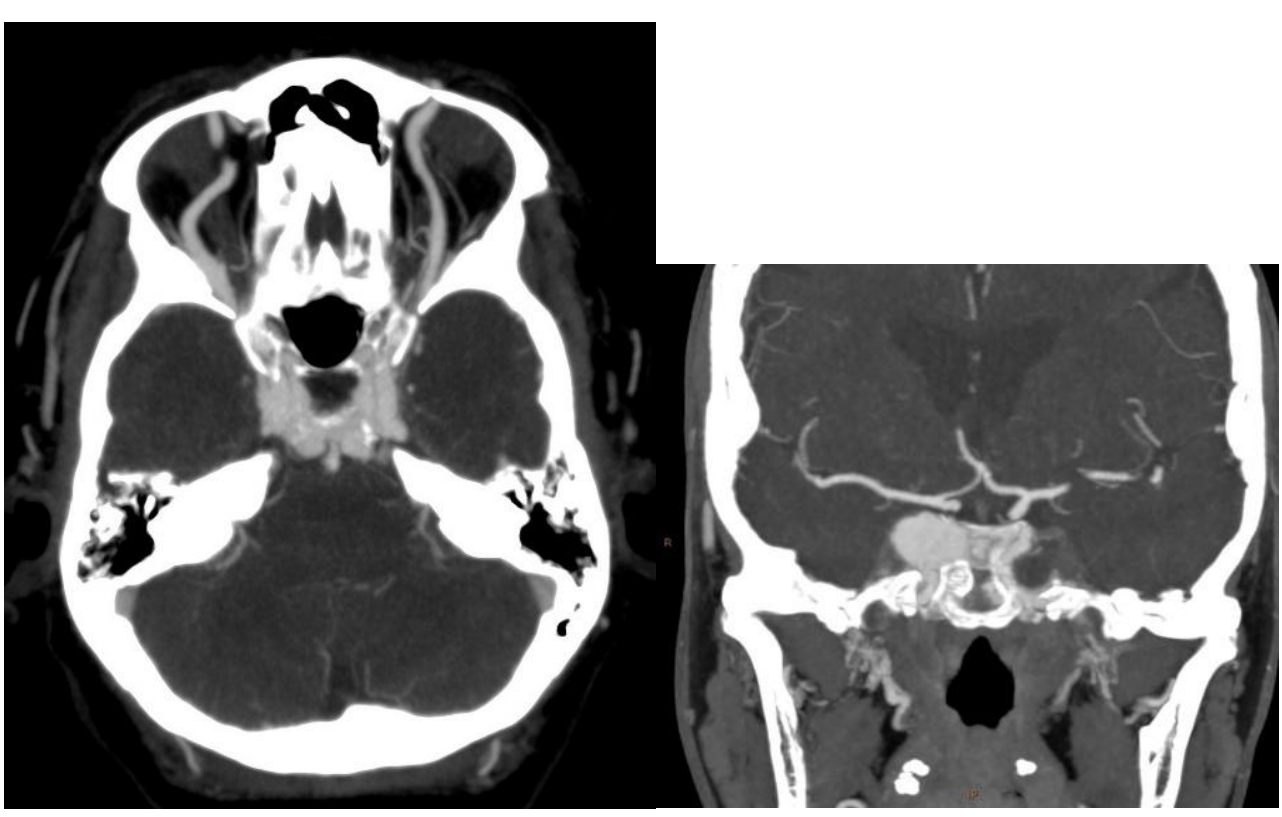
2. Self-audible **bruit** synchronous with pulse (**pulsatile tinnitus**);
   - many are also audible to examiner – at temple or orbit.
   - reduced by manual occlusion of carotid artery in neck (recession of exophthalmos may also be observed).

3. **Headache** (± other signs of ICP↑)

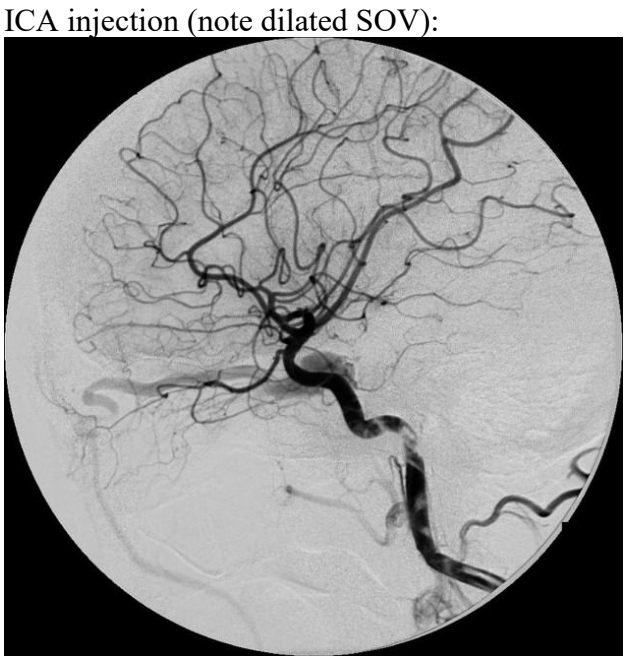
4. Exsanguinating **epistaxis** (H: place Foley into nose and hold manual carotid compression on the side of bleed while transporting to OR)

DIAGNOSIS

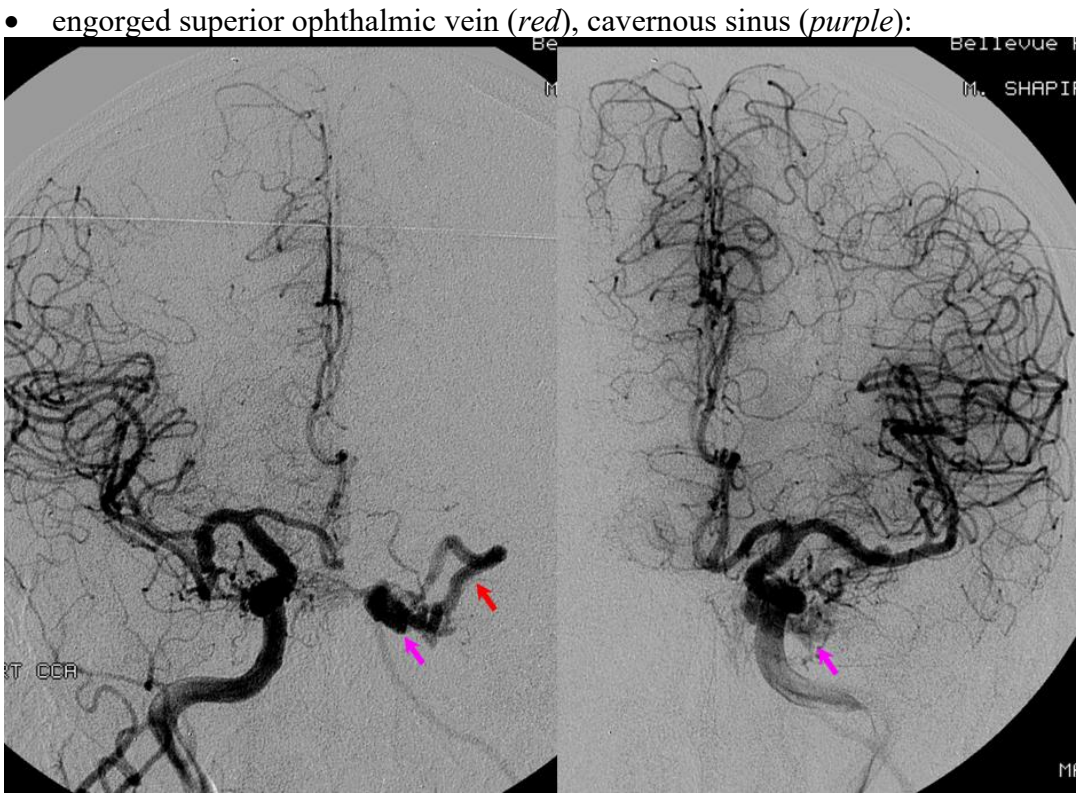
1. **CTA** (look for dilated ophthalmic veins*, bulging cavernous sinus, asymmetric enhancement of cavernous sinus with attenuation similar to that of internal carotid artery and higher than that of transverse sinus, contrast extravasation, intracranial hemorrhage from a ruptured cortical vein)
   *esp. superior ophthalmic vein (SOV)!
2. **Selective carotid ANGIOGRAPHY** (high-speed digital subtraction imaging in multiple views of both bilateral ICA and ECA*) - diagnostic test of choice (confirms diagnosis): early filling of cavernous sinus and its enlarged draining tributaries (esp. ophthalmic veins).
   *only for spontaneous fistulas
3. **Contrast CT of orbit** - proptosis, swelling of extraocular muscles, dilation of superior ophthalmic vein (→ enlarged superior orbital fissure), enlarged cavernous sinus.
4. **Orbital ultrasound** - findings as CT.
5. **Complete ophthalmologic workup**: visual acuity, funduscopy (direct and indirect), intraocular pressure & gonioscopy.
   Ask ophthalmologist to measure IOP!!!!



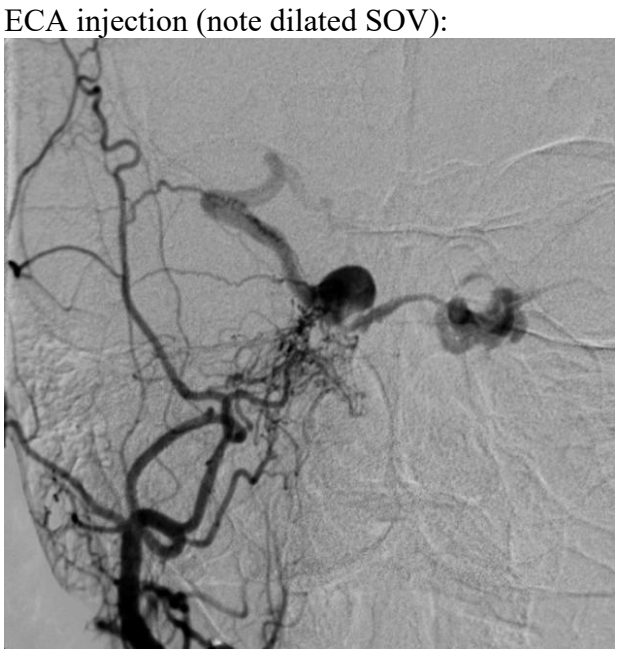
ICA injection (note dilated SOV):



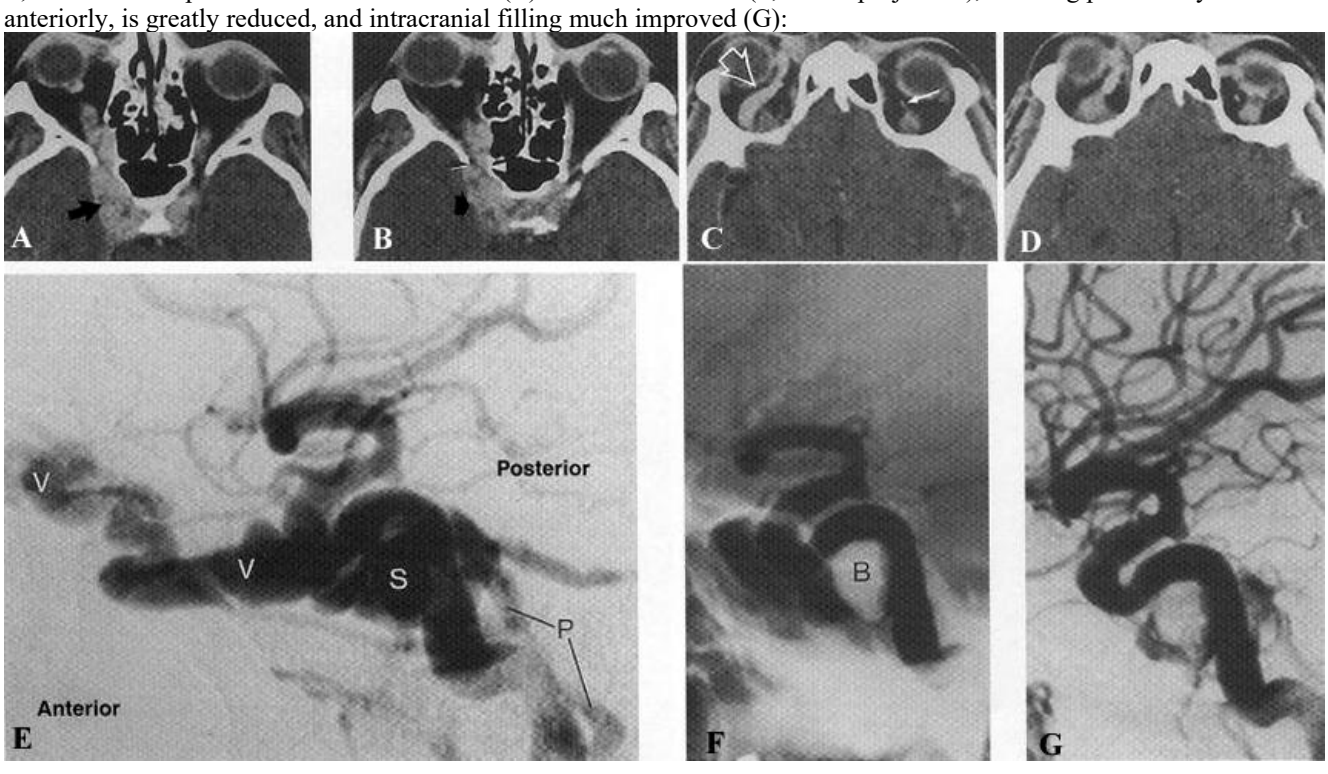
- engorged superior ophthalmic vein (*red*), cavernous sinus (*purple*):



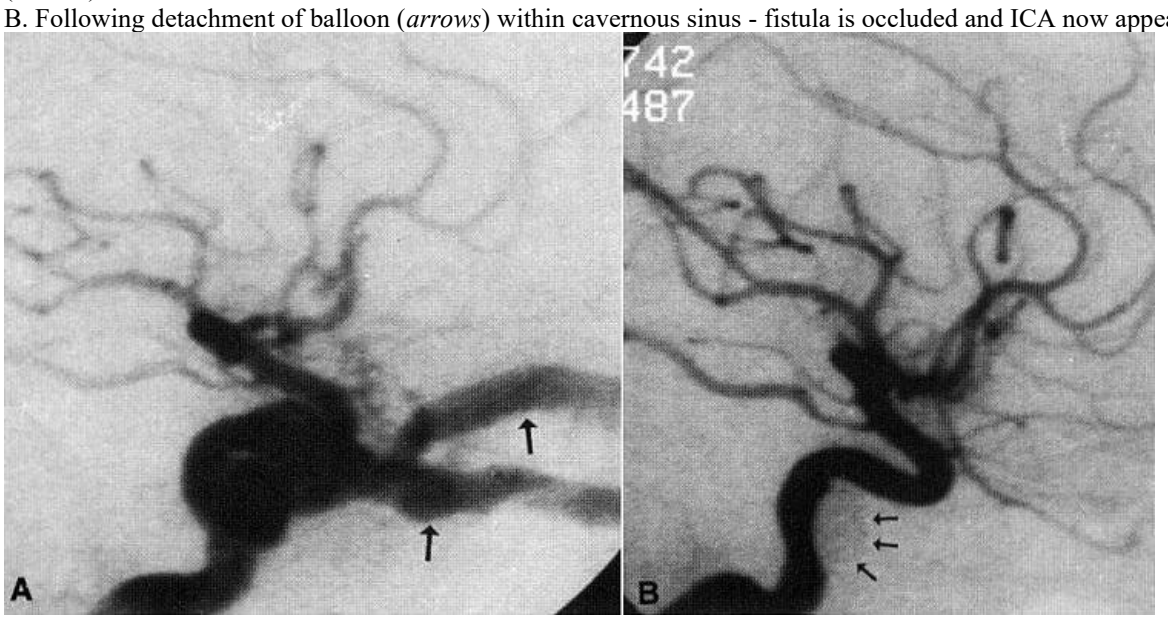
ECA injection (note dilated SOV):



A–D (axial contrast CT): right cavernous sinus (A, *thick black arrow*) is enlarged, and large enhancing mass runs forwards into orbit through widened superior orbital fissure (B, *arrowheads*); sigmoid structure (*open arrow* in C) in upper part of right orbit represents greatly dilated superior ophthalmic vein (cf. normal left side in C, *small white arrow*); some extraocular muscles are thicker than on left, and there is marked right proptosis.
E (right ICA intra-arterial DSA, lateral projection, arterial phase) - contrast medium floods into cavernous sinus (S), and drains forwards into grossly dilated superior ophthalmic vein (V); there is also shunting posteriorly and via inferior petrosal sinus (P); intracranial arterial filling is poor.
F, G - after therapeutic detachment of balloon (B) in cavernous sinus (F, lateral projection), shunting particularly anteriorly, is greatly reduced, and intracranial filling much improved (G):



A. Left ICA (lateral DSA) - rapid opacification of cavernous sinus and both superior and inferior ophthalmic veins (*arrows*).
B. Following detachment of balloon (*arrows*) within cavernous sinus - fistula is occluded and ICA now appears normal.



Carotid angiogram - large communication (*vertical arrow*) between ICA (above) and cavernous sinus; in addition to enlarged orbital veins that drain forward from cavernous sinus, there is backward drainage through petrosal sinus (*horizontal arrow*):

TREATMENT

Measure IOP – if > 20 → emergent treatment!
Cortical venous drainage → treat!
In acute setting of vision loss / CN paralysis, **glucocorticoids** (e.g. DEXAMETHASONE), DIAMOX may be used while waiting for definitive diagnosis and treatment.

Type-A fistulas rarely resolve spontaneously because of high flow (fistula enlarges, causing decreased chances of visual recovery).

- treatment indications - progressive visual loss (main complication!!!), intolerable bruit, cosmetic effects of proptosis.

Definitive management - **obliteration of fistulous connection** with preservation of ICA patency:

A. **Endovascular approach** - through arterial approach (N.B. ICA hole may be big – use balloon-assisted technique!)
   a) detachable coils – preferred method (pack cavernous sinus as much as you can) for simple and complex fistulas
   b) Onyx – if one simple cavity with one arterial feeder
   c) detachable balloon

d) ICA stenting (pipeline) across fistula may have role.

B. **Direct surgical exposure and obliteration** of fistula (now rarely indicated).

- symptoms & signs improve within days after treatment, but complete resolution may take weeks to months.
- severely refractory fistulas → surgical or endovascular **sacrifice of ICA** (+ clipping of supracavernous segment proximal to PComA to prevent fistula from stealing blood from cerebral vasculature).

Type B, C, D fistulas have higher incidence of *spontaneous resolution*.

- **carotid self-compression** for 20-30 seconds 4 times per hour may lead to fistula thrombosis.
  - patient is instructed to compress carotid artery on side of lesion using contralateral hand (should patient develop cerebral ischemia during compression, contralateral hand likely will be affected, releasing compression).
- if compression is not effective or if more rapid intervention is indicated → **endovascular fistula embolization**

  N.B. may prefer venous approach (posterior approach via *inferior petrosal sinus* or transocular via *superior ophthalmic vein**) *surgically expose vein to allow direct cannulation

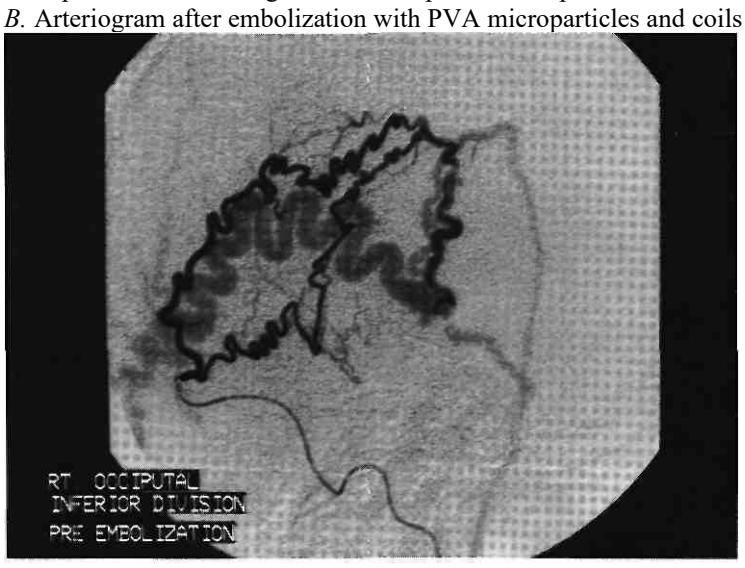
PROGNOSIS

- RECURRENCE rate 1-3.9%.
- routine ***follow-up angiogram*** - to ensure that fistula has not recurred or that alternate fistulous pathways have not developed. (H: second balloon treatment)

Other AV fistulae

- abnormal communications between artery and vein secondary to:
  a) most common - traumatic laceration of vessels (esp. GSW – routine CTA for all GSW patients on day 10-12; if retained bullet gives obscuring artefacts – then angiography)
  b) aneurysm
  c) angiodysplasia
- treated via **endovascular approach** (balloons, PVA, liquid agents, coils).

Traumatic AV fistula:
A. Superselective arteriogram of a. occipitalis - two prominent branches draining directly to markedly dilated draining vein.
B. Arteriogram after embolization with PVA microparticles and coils - nonfilling of draining vein.



(A)

(B)

BIBLIOGRAPHY for ch. "Head Trauma" → follow this LINK >>