

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

- 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).
- 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:

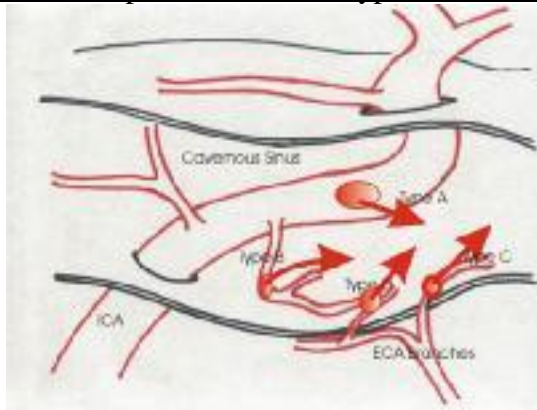
- low-flow.
- more common in **women > 50 years** (7:1 female-to-male ratio).
- most commonly **spontaneous** etiology.

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**:

- 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.
- progressive (over days or weeks) monocular **visual loss** in late stages
- cranial nerve palsy** (III, IV, V, VI) ipsilaterally or bilaterally
- dilatation of retinal veins, optic disc swelling, retinopathy.
- 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

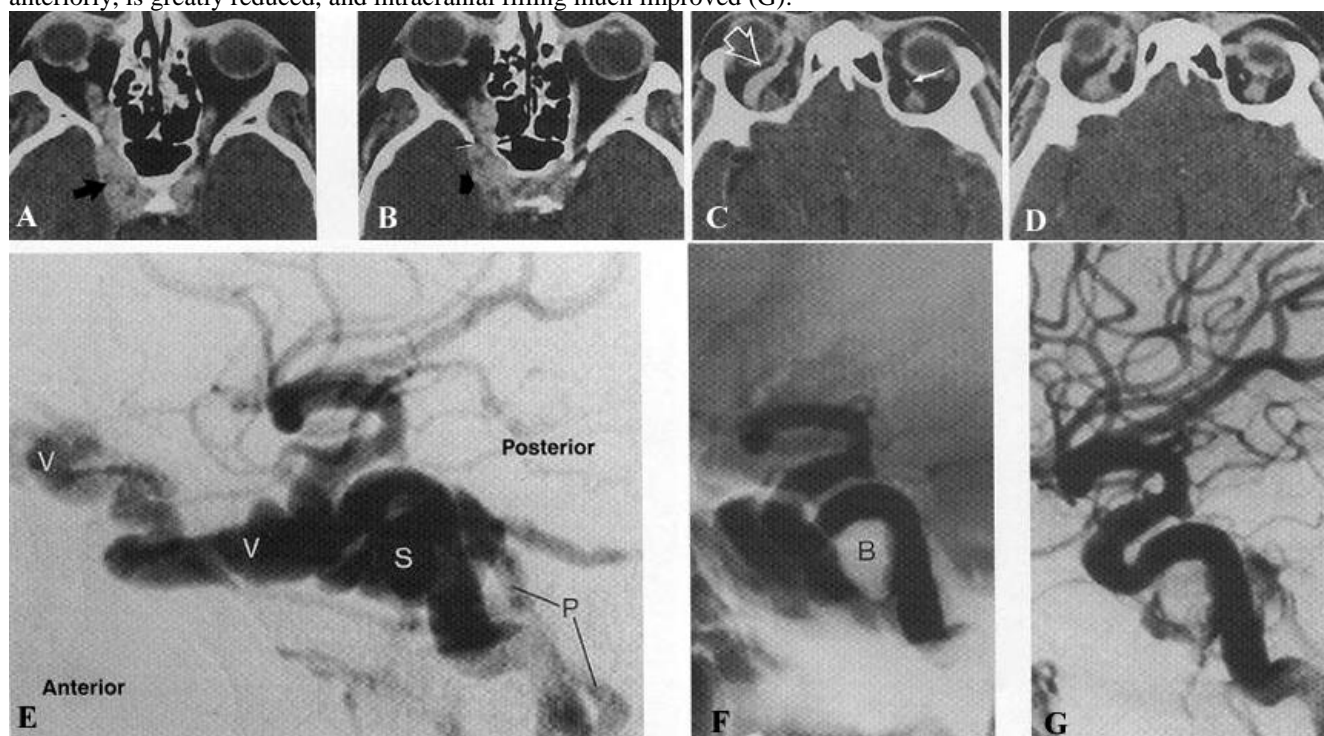
1. **CTA** (look for dilated ophthalmic veins, contrast extravasation).

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 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.

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.

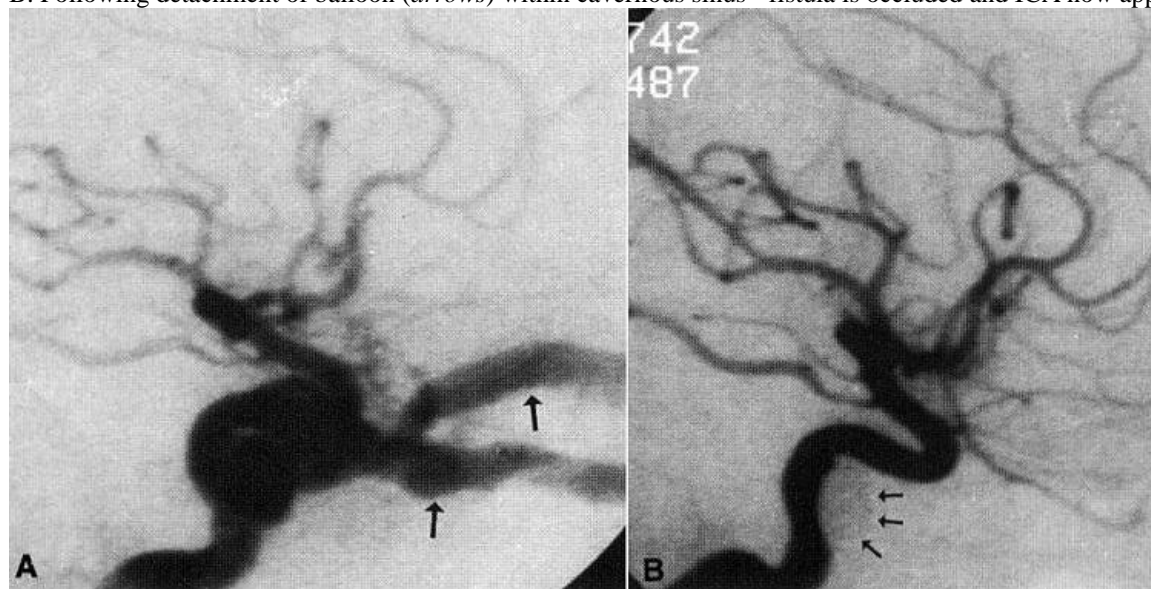
E (right ICA intra-arterial DSA, lateral projection, arterial phase) - contrast medium floods into cavernous sinus (S), and drains forwards into grossly dilated superior ophthalmic vein (V); there is also shunting posteriorly and via inferior petrosal sinus (P); intracranial arterial filling is poor.

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):

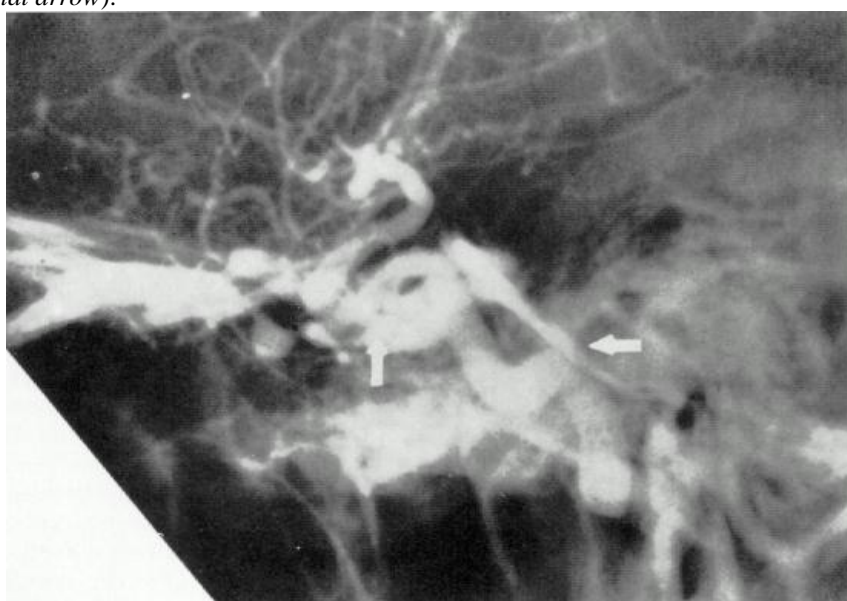


A. Left ICA (lateral DSA) - rapid opacification of cavernous sinus and both superior and inferior ophthalmic veins (*arrows*).

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

In acute setting of vision loss / CN paralysis, **glucocorticoids** (e.g. **DEXAMETHASONE**) 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 (ICA or ECA) or **venous** approach (inferior petrosal sinus, facial vein, superior ophthalmic vein*)
*surgically expose vein to allow direct cannulation
 - 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 in future.
- 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**

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:

- 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)
 - aneurysm**
 - angiodyplasia**
- 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 >>](#)