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).

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).

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:

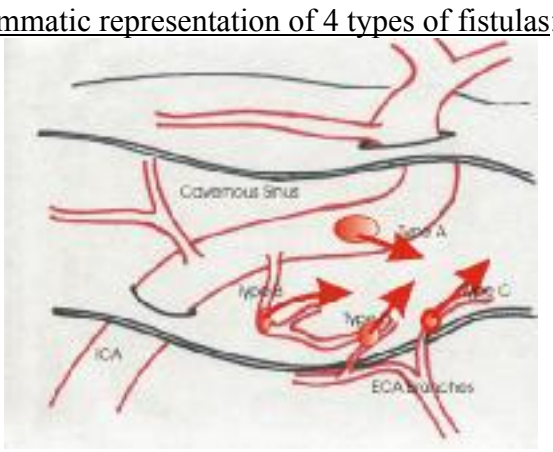
- low-flow.
- more common in *women > 50 years* (7:1 female-to-male ratio).
- most commonly spontaneous etiology.

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).

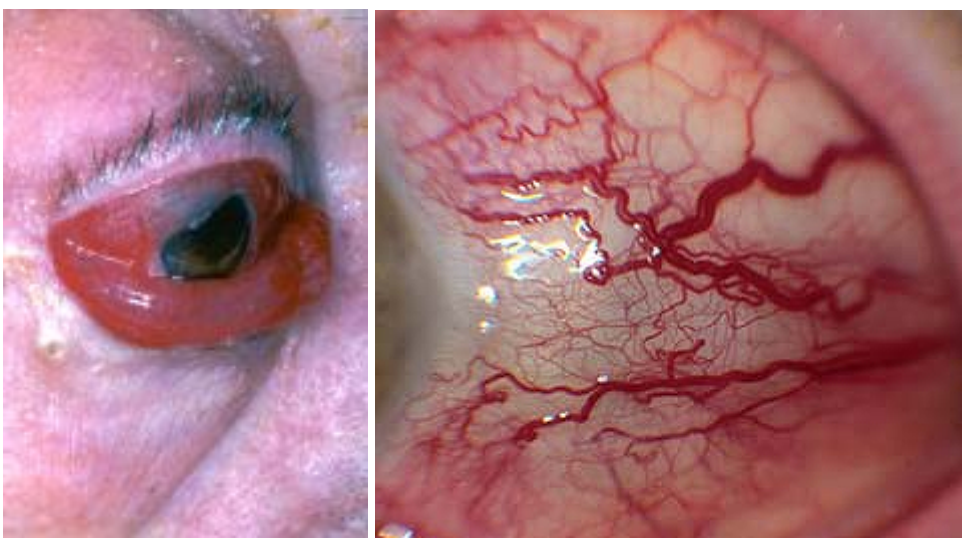
Diagrammatic representation of 4 types of fistulas:



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) progressive (over days or weeks) monocular visual loss in late stages
   3) cranial nerve palsy (III, IV, V, VI) ipsilaterally or bilaterally
   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;
   - 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↑)

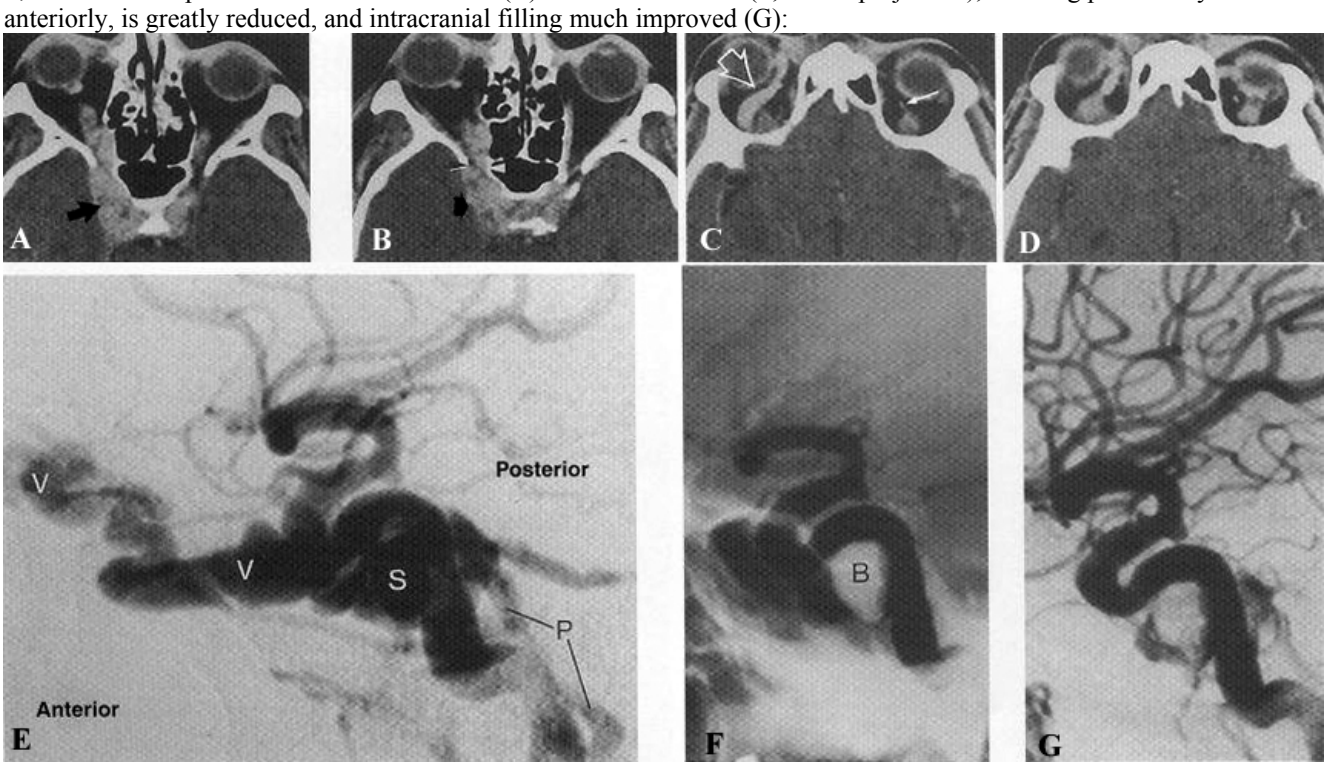
DIAGNOSIS

1. **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.

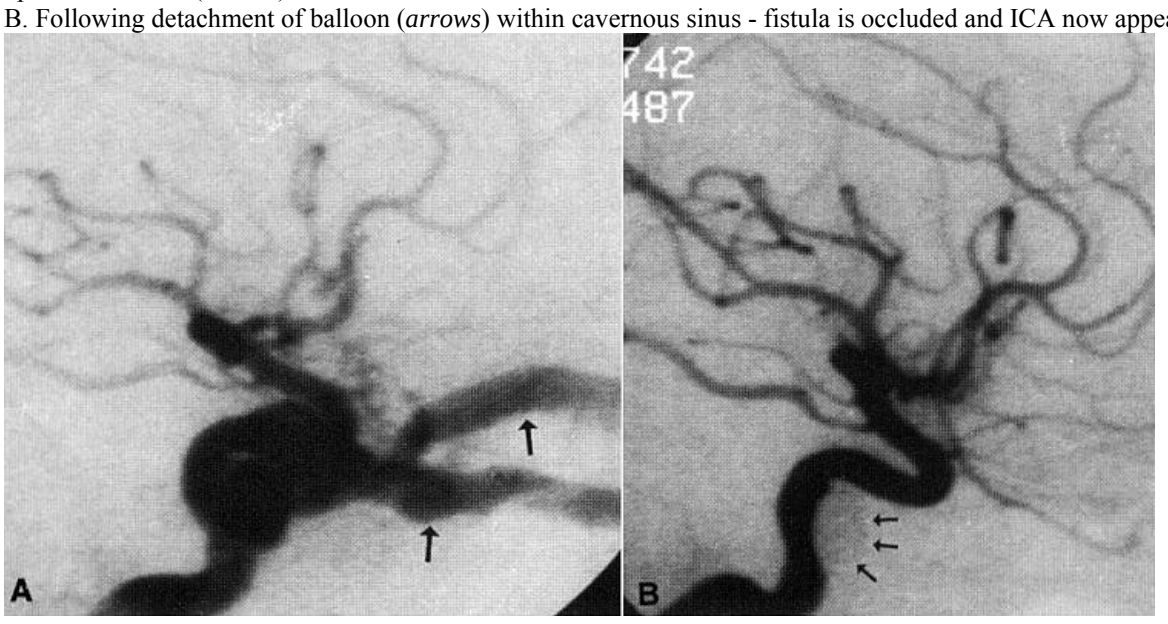
*only for spontaneous fistulas

2. **Contrast CT of orbit** - proptosis, swelling of extraocular muscles, dilation of superior ophthalmic vein (→ enlarged superior orbital fissure), enlarged cavernous sinus.
3. **Orbital ultrasound** - findings as CT.
4. **Complete ophthalmologic workup**: visual acuity, funduscopy (direct and indirect), intraocular pressure & gonioscopy.

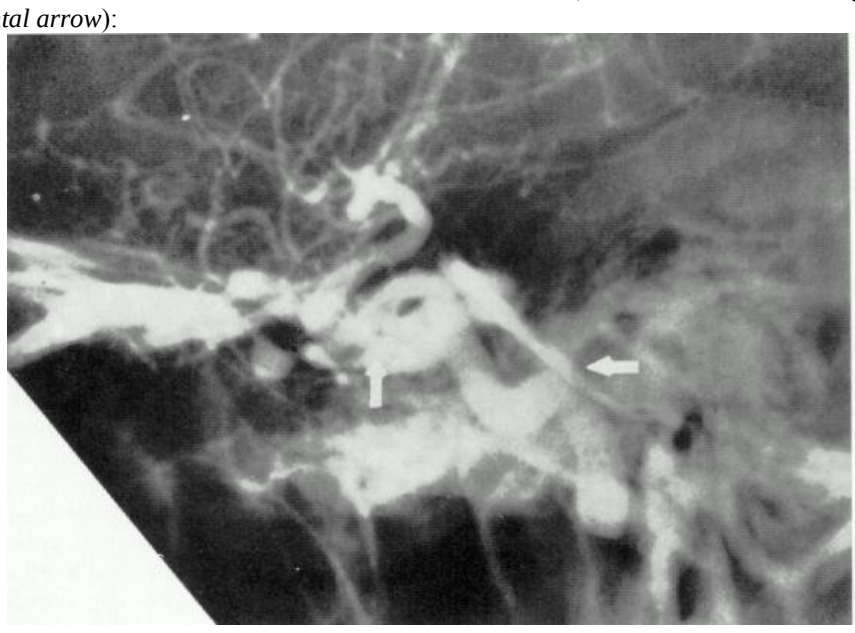
A–D (axial contrast CT): right cavernous sinus (A, *thick black arrow*) is enlarged, and large enhancing mass runs forwards into orbit through widened superior orbital fissure (B, *arrowheads*); sigmoid structure (*open arrow* in C) in upper part of right orbit represents greatly dilated superior ophthalmic 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 digital subtraction arteriogram - 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 (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 *ICA* or through *internal jugular vein* and petrosal sinus):
   a) detachable balloon [90-100% success]
   b) Guglielmi detachable coils
   c) ICA stenting 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 → selective **endovascular fistula embolization** through *ECA* (polyvinyl alcohol [PVA] is preferred embolic material).
  – occasionally, endovascular approach through *superior ophthalmic vein* is required (surgically expose vein to allow catheter placement).

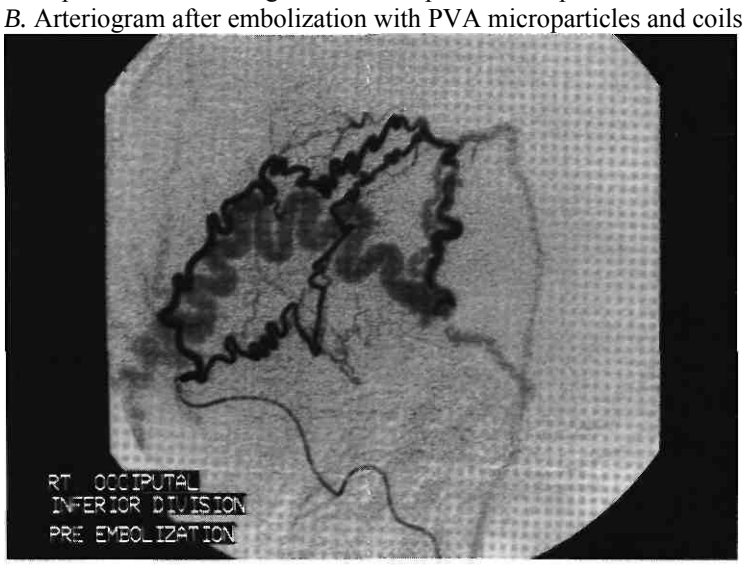
PROGNOSIS

- 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 (e.g. carotid-cavernous fistula)
  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 >>