Cerebral Arteriovenous (AV) Fistula

Forhad H. Chowdhury, Shamshul Alam, Nazmin Ahmed and Mohammod Raziul Haque

Abstract: A cerebral arteriovenous fistula (AVF) is an abnormal direct connection of intracranial arteries and vein without intervening capillaries. Cerebral AV fistulas include dural AVFs, caroticocavernous fistulas (CCFs), vein of Galen malformations and pial AVFs. The clinical presentation of a cerebral AVF varies according to the site and magnitude of the fistula. A CT scan, MRI, CTA, MRA and DSA are the necessary investigation methods for diagnosis, classification and management purposes. The mainstay of treatment is an endovascular occlusion. A microsurgical occlusion or excision is needed when the endovascular procedure fails or is not possible. Here, the pathology, classification, clinical presentation and management of dural AVFs, vein of Galen malformations and pial AVFs are discussed. CCFs will be discussed in Chapter 15.

Abbreviations

ACA	anterior cerebral artery	AV	Arteriovenous
AVF	arteriovenous fistula	CT	computed tomography
CTA	computed tomographic angiogram	CVR	cortical venous reflux
DAVF/dAVF	dural arteriovenous fistula	DSA	digital subtraction angiogram
ECA	external carotid artery	ICA	internal carotid artery
ICH	intracranial hematoma	MHT	meningohypophyseal trunk
MMA	middle meningeal artery	MRI	magnetic resonance imaging
MRA	magnetic resonance angiogram	NBCA	n-butyl cyanoacrylate
PCA	posterior cerebral artery	PICA	posterior inferior cerebellar artery
SCA	superior cerebellar artery	SPS	superior petrosal sinus
SS	sigmoid sinus	SSS	superior sagittal sinus
TS	transverse sinus	VOG	vein of Galen
PAVF	pial arteriovenous angiogram		

1. Introduction

An arteriovenous fistula (AVF) is a pathological connection of vessels in the brain or the spinal cord, where one or more arteries directly drain into one or more veins or venous sinuses (Figure 1). If a fistula is formed, there is communication between the intracranial artery and vein that leads the venous system to become arterialized, resulting in a mass effect or brain hemorrhage (Gupta and Periakaruppan 2009; Reynolds et al. 2017).

Cerebral arteriovenous (AV) fistulas are discussed here in three groups:

1. Dural AV fistula;
2. Vein of Galen (VOG) malformation;
3. Pial AV fistula.

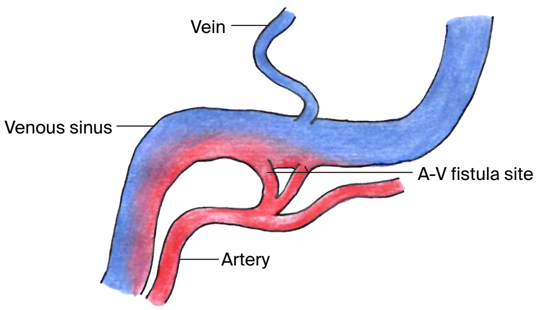

Figure 1. Schematic diagram of the arteriovenous fistula. Source: Figure by authors.

2. Dural AV Fistula (DAVF)

2.1. Epidemiology

Of intracranial vascular malformations, 10–15% are DAVFs (Luciani et al. 2001), typically encountered in middle-aged adults (acquired disease).

2.2. Distribution of Dural Supply

Group A: entire cerebral convexity, lateral segment of the cerebellar convexity and falx cerebri, supplied by the middle meningeal artery (MMA) convexity branch.

Group B: the cranial base perfused by external as well as internal carotid arteries.

Group C: the medial cerebellar convexity supplied by the vertebrobasilar system (mainly posterior meningeal).

The incidence of DAVFs at various locations is as follows (Lasjaunias et al. 1986):

- Transverse sinus (TS), 50% of cases;
- Cavernous sinus (CS), 16% of cases;
- Tentorium cerebelli, 12% of cases;
- Superior sagittal sinus (SSS), 8% of cases.

2.3. Classification of DAVFs

Before classification, the diagnosis and all information are collected by neuro-imaging. The essential modalities of investigations are a CT scan with a CTA, MRI with an MRA and gold standard cerebral DSA.

2.3.1. Cognard Classification

I	Antegrade meningeal vein or dural venous sinus
IIa	Retrograde into the dural venous sinus/meningeal vein
IIb	Antegrade and cortical venous reflux (CVR) (10–20% hemorrhage)
III	CVR no ectasia (40% hemorrhage)
IV	CVR with ectasia (65% hemorrhage)
V	Spinal venous drainage (Cognard et al. 1995)

2.3.2. Simplified Borden Classification

Lesion type	Definition
I	Draining is directly anterograde to the major sinus of the vein
II	Drains to the vein sinus, then back to the subarachnoid veins via retrograde drainage
III	Drains directly to subarachnoid veins (Figure 2) (Borden et al. 1995; Nader et al. 2014)

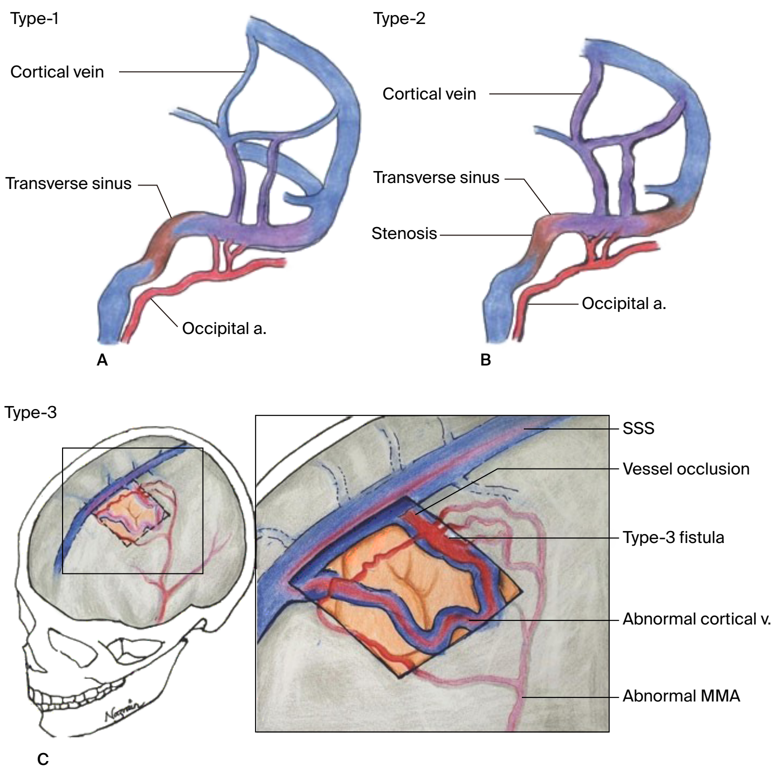

Figure 2. (**A**–**C**) Schematic sequential overview of the Borden classification of DAVFs. Source: Figure by authors.

2.4. Signs and Symptoms

People with DAVFs may experience no symptoms at all. When symptoms are present, they might range from minor discomfort to serious hemorrhage.

- Tinnitus (pulsatile)
- Headache
- Visual impairment
- Bruit (sound heard due to unusual blood flow)
- Seizure (Gupta and Periakaruppan 2009; Reynolds et al. 2017)

DAVF presentation is defined by its site, as well as pattern of venous drainage (Tables 1 and 2).

Table 1. Clinical features according to anatomical type of DAVF.

DAVF	Clinical Features
Transverse–sigmoid sinus DAVFs	Pulsatile tinnitus.
Cavernous sinus DAVFs	Exophthalmos, chemosis and blindness.
Superior sagittal sinus DAVFs	Hemorrhage, local venous congestion, cerebral edema and cerebral ischemia.
DAVFs with a premedullary draining vein	Myelopathy and progressive tetraplegia.

Source: Authors' compilation based on data from Gupta and Periakaruppan (2009), Reynolds et al. (2017) and Greenberg (2010).

Table 2. Clinical manifestations of DAVFs.

Clinical Features
Clinical features of an intracerebral hemorrhage (ICH)
Focal neurological deficit (such as limb weakness, dysphasia, cerebellar features, myelopathy) Generalized neurological deficit (such as dementia)
Tinnitus (pulsatile) and bruit (Objective)
Chemosis, conjunctival injection, proptosis, ophthalmoplegia
Visual impairment (due to increased intraocular pressure, orbital congestion, optic neuropathy or retinal hemorrhages)
Papilledema (due to pseudotumor cerebri or hydrocephalus induced by disturbed venous drainage)
Glaucoma
Facial pain (for compression of the 1st and 2nd division of the 5th nerve in the lateral wall of the cavernous sinus)
Headache
Pulsatile mass (palpated behind the mastoid process in the course of the occipital artery), seizure (focal or generalized)

Source: Authors' compilation based on data from Gupta and Periakaruppan (2009), Reynolds et al. (2017) and Greenberg (2010).

2.5. Risk Factors

Those predisposed to vein thrombosis, such as coagulation disorders that enhance the probability of a venous sinus occlusion, have genetic risk factors for DAVFs.

Persons in their late middle age (50–60 years) are the most commonly affected by DAVFs. DAVFs, on the other hand, can affect people of all ages, including youngsters.

Benign meningeal tumors have been linked to the development of DAVFs according to new research (Gupta and Periakaruppan 2009; Reynolds et al. 2017; Greenberg 2010).

2.6. Treatment of Arteriovenous Fistulas (DAVFs)

- Endovascular embolization—Embolization is the choice of treatment for most DAVFs. During this surgery, the neurosurgeon inserts a catheter into the arteries leading to the DAVF in the brain and injects liquid embolizing agents like NBCA, Onyx or glue into the feeding arteries. The injection blocks that artery, reducing blood flow via the DAVF.

- Microsurgical excision—This is used to close DAVFs that are not closed with endovascular treatment. We perform a craniotomy and separate the DAVF from the tissues surrounding the brain/spinal cord utilizing microsurgical techniques.

2.6.1. Surgical Technique

Two general approaches can be considered surgically. The first is the disconnection of the retrograde cortical venous drainage from the DAVF, leaving the DAVF intact but without any cortical venous reflux. This removes the danger from hemorrhage. The second approach is to remove the DAVF itself. In this procedure, it is critical that the sinus resection does not impair the normal cortical venous drainage.

1. Disconnection of cortical venous drainage: This is applied for all locations providing retrograde flow. The smallest approach possible with a clip is applied as close to the entry of the vein to the dura as possible.
2. Venous sinus resection involved in the DAVF: This approach is rarely required due to the introduction of liquid embolic agents and intravenous sinus temporary balloon protection of the venous sinus lumen. If the involved venous sinus is no longer functioning with normal venous drainage (thrombosed sinus, no important tributaries from the brain with collateral venous drainage exists), the DAVF with the venous sinus can be excised (Gupta and Periakaruppan 2009; Reynolds et al. 2017).

2.6.2. Endovascular Treatment

An Onyx trans-arterial embolization (TAE) is simple and effective therapy for DAVFs.

MMAs are the most feasible targets of Onyx TAEs except for skull base DAVFs.

Excessive Onyx migration into intact veins, as well as dangerous anastomoses among dural arteries should be avoided (Gupta and Periakaruppan 2009; Reynolds et al. 2017).

2.6.3. Gama-Knife Surgery

Gamma-knife surgery may be utilized to attack and shut the fistula. Radiation therapies take time to work (months–years), but they can be extremely effective at closing exceedingly difficult fistulas that have no other therapy options and when embolization or surgery is not possible.

2.7. Types of AV Fistulas (According to the Location) and Their Surgical Approach

a. **Transverse sinus and sigmoid sinus (SS) DAVFs:** The TS and SS have abnormal connections with several dural arterial branches arising from the MMA and the transmastoid branch originating from the occipital artery. A lateral suboccipital approach can be used to reach and treat it (Nader et al. 2014).
b. **Superior petrosal sinus AV fistula:** These can be exposed by a retro-sigmoid approach. Superior petrosal sinus (SPS) DAVFs are a subgroup of tentorial DAVFs that are situated at the petro-tentorial junction (Figures 3 and 4) and get internal carotid artery (ICA) perfusion from the meningohypophyseal trunk (MHT) and venous drainage into the petrosal vein, Rosenthal vein, lateral mesencephalic vein or cerebellar hemispheric veins (Luciani et al. 2001).

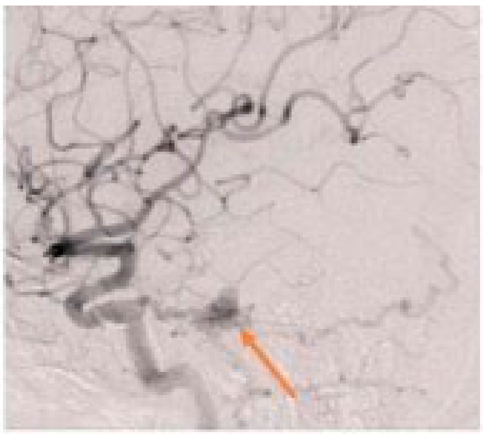

Figure 3. Superior petrosal sinus DAVF. Source: Figure by authors.

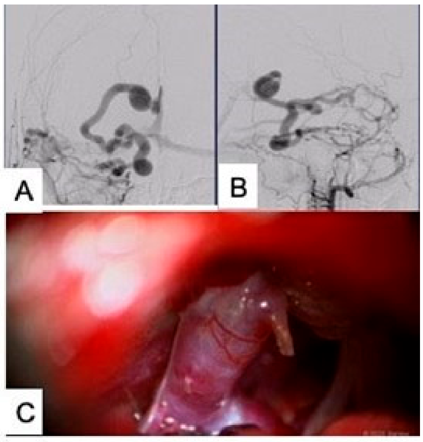

Figure 4. (**A**,**B**) DSA showing a right-sided superior petrosal sinus dural AVF. (**C**) Right extended retrosigmoid craniotomy and clipping of a superior petrosal sinus dural AVF. Source: Figure by authors.

c. **Middle fossa and cavernous sinus DAVFs:** These can be exposed via the middle fossa approach or lateral supraorbital approach.

d. **Tentorial DAVFs (Figure 5):** The arterial supply demonstrated on DSA is usually complex, multiple and in combination form. The feeders are from the following:

- ICA through tentorial branches of the ICA
  - The MHT (usually);
  - The inferolateral trunk.
- External carotid artery (ECA)
  - The MMA (usually);
  - The occipital artery (commonly);
  - The ascending pharyngeal artery.
- Vertebral artery
  - The musculospiral (extradural) artery;
  - The posterior meningeal artery.
- Basilar artery
  - The posterior inferior cerebellar artery (PICA);
  - Anterior inferior cerebellar artery;
  - Medial dural–tentorial branch of the superior cerebellar artery (SCA) (previously underrecognized);
  - The tentorial branch of the posterior cerebral artery (also known as the artery of Davidoff and Schechter).

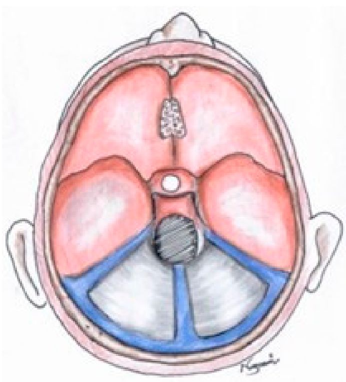

Figure 5. Location of different types of tentorial DAVFs such as-1: falx cerebellum, 2: torcular, and 3: petrotentorial on the right or left. Source: Figure by authors.

In these patients, a flow-related aneurysm can arise from a feeding PICA. There may be a bilateral fistulous supply, mainly involving the posterior division of the MMA and occipital arteries (Byrne and Garcia 2013; Nader et al. 2014).

Approach: The approach depends on the location. It can be exposed through the subtemporal approach in a park bench position.

e. Frontobasal DAVFs (Figures 6 and 7): Feeders are from the following:

- Bilateral ophthalmic arteries
    - Anterior ethmoidal arteries;
    - Posterior ethmoidal arteries.
- Bilateral external carotid arteries
    - From the ethmoidal and cavity branches of the internal maxillary arteries.
- Anterior cerebral arteries (ACA) (rare)
    - Frontopolar artery;
    - Orbitofrontal artery.

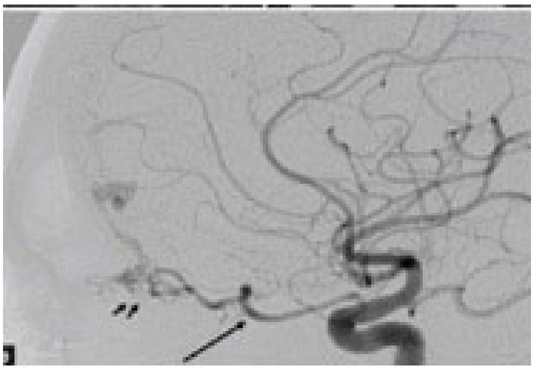

Figure 6. Frontobasal DAVF. Source: Figure by authors.

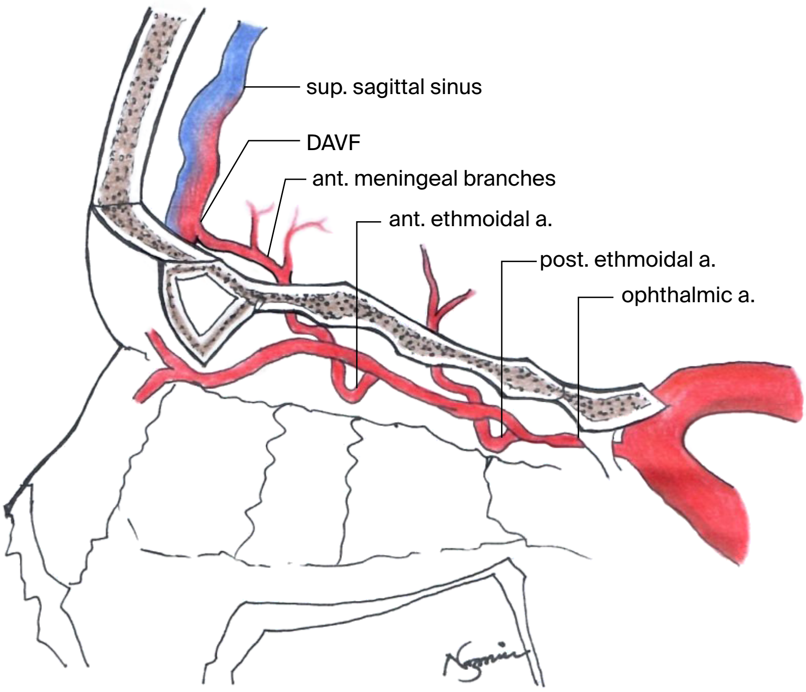

Figure 7. Diagram of a superior sagittal sinus fistula drained by feeders of the ophthalmic artery that was treated via the endoscopic endonasal approach for obliteration. Relevant branches of the ophthalmic artery are displayed. Source: Figure by authors.

Venous drainage is usually occurs in the rostral part of the SSS. It can be exposed through the anterior inter-hemispheric approach.

f. **Superior sagittal sinus DAVFs**: Arterial supply comes from the MMA. It can be approached through the midline frontal or parietal craniotomy approach (Figure 8).

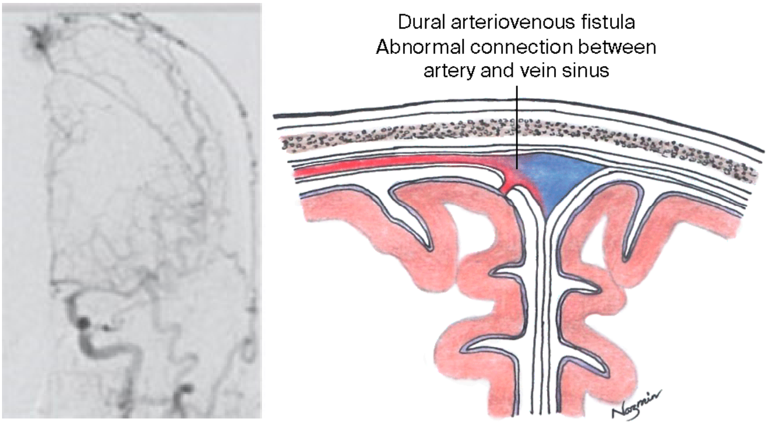

Figure 8. Superior sagittal sinus DAVF. Source: Figure by authors.

g. **Posterior fossa dural AV fistula**: Its arterial supply comes from the posterior meningeal artery, MMA and occipital artery. It can be approached by suboccipital craniotomy (Figure 9).

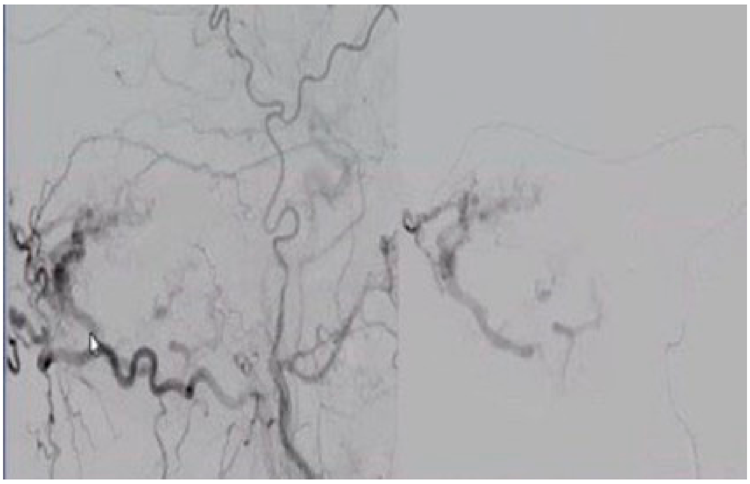

Figure 9. Posterior fossa DAVF. Source: Figure by authors.

h. **Spinal AV fistula** (also discussed in Chapter 16): When a fistula is formed, blood from an artery with high pressure flows directly into a low-pressure vein of low-flow anatomical structure. Hence, it causes venous congestion and spinal cord swelling. As the spinal cord swells, most patients begin to experience leg paralysis, back and leg pain, and bladder and bowel abnormalities. The patient may feel stiffness in the legs, shakiness or any other descriptive adjectives depending on what type of weakness (upper vs. lower motor neuron or both). The symptoms of bowel and bladder disorders might be vague: a person may feel unable to begin peeing, they may feel unable to control their bladder as they should or they may sense the urge to urinate and thus have incontinence. Fecal incontinence or constipation are also possible side effects. Male patients can also have impotency.

2.7.1. Diagnosis

1. Carried out by a spinal digital subtraction angiogram (DSA);
2. An MRI scan does not demonstrate the fistula site but shows the evidence of signal changes and probable location of the fistula, and a spinal MRA can show the fistula;
3. A spinal CTA is very helpful for surgery.

2.7.2. Treatment

The aim of management is to shut the fistula either by plugging it with endovascular embolization or by microsurgical disconnection by laminectomy.

It is approached by laminectomy and resection of the fistula.

2.7.3. Complication of Microsurgery

- Sever bleeding: >1000 mL;
- Ischemic complication: postoperative infarction;
- Hemorrhagic complication: ICH, epidural hematoma.

3. Vein of Galen Malformation

3.1. Introduction

A VOG malformation is a congenital abnormal formation of cerebral vessels that happens before birth. In this malformation, blood flows directly from cerebral arteries to a dilated great cerebral VOG.

The pressure, either directly from an artery through an AVF or by a tributary vein that gets flow directly from an artery, causes enlargement of the VOG.

3.2. Clinical Features

In neonates, malformations frequently cause heart failure, hydrocephalus and cranial bruits, as well as subarachnoid hemorrhage. Cardiac failure is caused by a magnitude of the arteriovenous shunt, which can steal up to 80% of the cardiac output by returning huge amount of blood under high pressure to the right side of heart, as well as pulmonary circulation, and the presence of a sinus venosus atrial septal defect. It is also the leading mode of death in these patients.

3.3. Classifications

Four types of vein of Galen malformations have been demonstrated:

- **Type I:** small pure cisternal fistula between the vein of Galen (voG) and either the pericallosal arteries (anterior or posterior) or posterior cerebral artery
- **Type II:** multiple fistulous communications between the vein of Galen and the thalamoperforating vessels
- **Type III:** high flow mixed type I and II
- **Type IV:** parenchymal arteriovenous malformation (AVM) with drainage into the vein of Galen
    - IVA: thalamic AVM
    - IVB: mesencephalic AVM
    - IVC: mesodiencephalic and cisternal AVM

3.4. Treatment: Options

a. Microsurgery;
b. Endovascular treatment by:
    - the arterial route or
    - venous route.

3.5. Outcomes

Babies often die either during the neonatal period or early infancy. In most patients, a VOG malformation cannot be corrected. Most die from an intracranial hemorrhage (Nader et al. 2014; Greenberg 2010; Gupta and Varma 2004).

4. Pial AV Fistula

Pial arteriovenous fistulas (PAVFs) are a type of cerebral vascular malformation that accounts for just 1.6% of all intracranial vascular abnormalities (Alurkar et al. 2016; Halbach et al. 1989). A brain AV venous malformation, on the other hand, has a nidus between the artery and the draining vein. Because there is no tangle of veins between the artery and the vein in a pial fistula, there is a greater pressure gradient, making the lesion more sensitive to rupture and causing symptoms, resulting in a worse prognosis for individuals with a PAVF. It usually presents with hemorrhage and rarely with pressure symptoms. The essential modalities of investigations are a CT scan with a CTA, MRI with an MRA and gold standard cerebral DSA (Alurkar et al. 2016).

Because of the high mortality rate associated with untreated instances, these lesions should be carefully diagnosed and treated as soon as possible, either via endovascular or microsurgical means (Alurkar et al. 2016; Nelson et al. 1992).

Author Contributions: Conceptualization, methodology, validation, formal analysis, investigation, resources, data curation, F.H.C., S.A. and N.A.; writing—original draft preparation, F.H.C. and S.A.; writing—review and editing, F.H.C.; visualization, supervision, N.A. and M.R.H. All authors have read and agreed to the published version of the manuscript.

Funding: This research received no external funding.

Conflicts of Interest: The authors declare no conflicts of interest.

References

Alurkar, Anand, Lakshmi Sudha Prasanna Karanam, Suresh Nayak, and Rajesh Kumar Ghanta. 2016. Intracranial Pial Arteriovenous Fistulae: Diagnosis and Treatment Techniques in Pediatric Patients with Review of Literature. *Journal of Clinical Imaging Science* 6: 2. [CrossRef] [PubMed]

Borden, Jonathan A., Julian K. Wu, and William A. Shucart. 1995. A proposed classification for spinal and cranial dural arteriovenous fistulous malformations and implications for treatment. *Journal of Neurosurgery* 82: 166–79. [CrossRef] [PubMed]

Byrne, J. V., and M. Garcia. 2013. Tentorial Dural Fistulas: Endovascular Management and Description of the Medial Dural-Tentorial Branch of the Superior Cerebellar Artery. *American Journal of Neuroradiology* 34: 1798–804. [CrossRef] [PubMed]

Cognard, Christophe, Yves P. Gobin, Laurent Pierot, Anne-Laure Bailly, Emmanuel Houdart, Alfredo Casasco, Jacques Chiras, and Jean-Jacques Merland. 1995. Cerebral dural arteriovenous fistulas: Clinical and angiographic correlation with a revised classification of venous drainage. *Radiology* 194: 671–80. [CrossRef] [PubMed]

Greenberg, Mark S. 2010. *Handbook of Neurosurgery*. New York: Thieme.

Gupta, A. K., and A. L. Periakaruppan. 2009. Intracranial dural arteriovenous fistulas: A Review. *Indian Journal of Radiology and Imaging* 19: 43–48. [CrossRef] [PubMed]

Gupta, A. K., and D. R. Varma. 2004. Vein of Galen malformations: Review. *Neurology India* 52: 43–53. [PubMed]

Halbach, Van V., Randall T. Higashida, Grant B. Hieshima, Carl W. Hardin, Christopher F. Dowd, and Stanley L. Barnwell. 1989. Transarterial occlusion of solitary intracerebral arteriovenous fistulas. *American Journal of Neuroradiology* 10: 747–52. [PubMed]

Lasjaunias, Pierre, Ming Chiu, Karel Ter Brugge, Atul Tolia, Michel Hurth, and Mark Bernstein. 1986. Neurological manifestations of intracranial dural arteriovenous malformations. *Journal of Neurosurgery* 64: 724–30. [CrossRef] [PubMed]

Luciani, Alain, Emmanuel Houdart, Charbel Mounayer, Jean-Pierre Saint Maurice, and Jean-Jacques Merland. 2001. Spontaneous closure of dural arteriovenous fistulas: Report of three cases and review of the literature. *American Journal of Neuroradiology* 22: 992–96. [PubMed]

Nader, Remi, Cristian Gragnaniello, Scott C. Berta, Abdulrahman J. Sabbagh, and Michael L. Levy. 2014. *Neurosurgery Tricks of the Trade-Cranial: Cranial*. New York: Thieme.

Nelson, K., Y. Nimi, P. Lasjaunias, and A. Berenstein. 1992. Endovascular embolization of congenital intracranial pial arteriovenous fistulas. *Neuroimaging Clinics of North America* 2: 309–17.

Reynolds, Matthew R., Giuseppe Lanzino, and Gregory J. Zipfel. 2017. Intracranial Dural Arteriovenous Fistulae. *Stroke* 48: 1424–31. [CrossRef] [PubMed]

Caroticocavernous Fistula (CCF)

Forhad H. Chowdhury, Shamshul Alam, Nazmin Ahmed and Mohammod Raziul Haque

Abstract: A carotid cavernous fistula (CCF) is a relatively common intracranial arteriovenous fistula, usually traumatic and spontaneous in origin. A CCF may be of direct or indirect variety, and clinically presents with pulsatile proptosis, chemosis and red eye. Investigations include a CT of the head, an MRI of the head, and a CTA, MRA and DSA of the brain. Endovascular therapy comes first in line, but microsurgical treatment is needed where endovascular therapy is not possible or fails. Here, the etiopathogenesis, classification and management of a CCF are discussed concisely.

Abbreviations

AV	Arteriovenous	CCF	caroticocavernous fistula
CT	computed tomography	CTA	computed tomographic angiogram
CS	cavernous sinus	DAVF	dural arteriovenous fistula
DSA	digital subtraction angiography	ECA	external carotid artery
ICA	internal carotid artery	IPS	inferior petrosal sinus
MRA	magnetic resonance angiogram	MRI	magnetic resonance imaging
RTA	road traffic accident		

1. Introduction

A carotid cavernous fistula (CCF) is commonly traumatic in origin, following road traffic accidents (RTAs) due to the avulsion or tear of the cavernous carotid artery. Hence, blood goes to the cavernous sinus (CS) of the same side, followed by the opposite CS and the superior ophthalmic vein (Greenberg 2010).

2. Anatomy of the Cavernous Sinus

The CS is a complicated venous region that runs from the sphenoid bone to the periosteal, as well as the meningeal layers of the dura. On both sides of the sellae turcica, there is a twin venous space. The superior and inferior intercavernous sinuses provide open communication between the two regions. From the superior orbital fissure (SOF) to the petrous apex, the CS extends anteriorly. The diaphragm sellae are located cranially, and the larger wing of the sphenoid is located caudally. The dura forms a lateral boundary for the sinus (Figure 1). The ICA, with its periarterial sympathetic plexus, is located medially within the CS, and its involvement may produce Horner's syndrome. The sixth nerve is located to the lateral of the ICA. The ophthalmic and maxillary divisions of the trigeminal nerve, oculomotor nerve and trochlear nerve are located within the lateral dural boundary of the CS. Chemosis and proptosis can occur when venous drainage is compromised. The venous linkages in the CS are complicated and valveless. It communicates with practically every key venous component in the head and neck, either directly or indirectly. The superior and inferior ophthalmic veins, the sphenoparietal sinuses, and the middle meningeal vein all drain the CS. The superior and inferior ophthalmic veins connect the CS to the facial vein and pterygoid venous plexus. The CS drains into the superior and inferior petrosal sinuses, which then flow into the sigmoid sinus and internal jugular vein (Chowdhury et al. 2012; Tang et al. 2010; Barrow et al. 1985).

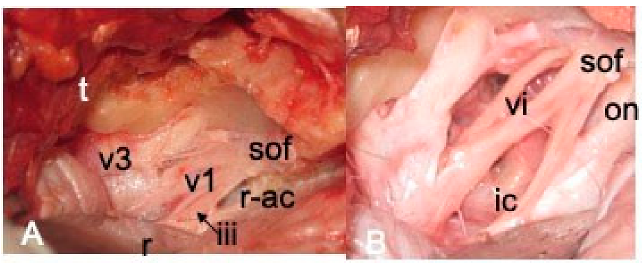

Figure 1. (**A**,**B**) Cadaveric dissection of the left CS after peeling of the dura from the CS with retraction of the temporal lobe extraduraly. (r—retractor, t—temporalis muscle, V3—mandibular nerve, V1—ophthalmic nerve, sof—superior orbital fissure, iii—oculomotor nerve, r-ac—root of anterior clinoid process (after drilling), vi—abducent nerve, ic—internal carotid artery and on—optic nerve). Source: Figure by authors.

3. Classification of the CCF

The CCF is generally classified depending on the arterial supply (Table 1, Figure 2). The clinical features and management approach are, however, mainly based on venous drainage.

Table 1. Barrow Classification.

Type	Pathogenesis	Arterial Supply	Hemodynamics
A	Head trauma/aneurysm rupture	ICA	High flow
B	Spontaneous	Dural branches of the ICA	Low flow
C	Spontaneous	Dural branches of the ECA	Low flow
D	Spontaneous	Dural branches of the ICA and ECA	Low flow

ICA: internal carotid artery, ECA: external carotid artery. Source: Authors' compilation based on data from Barrow et al. 1985; Cruz 1998.

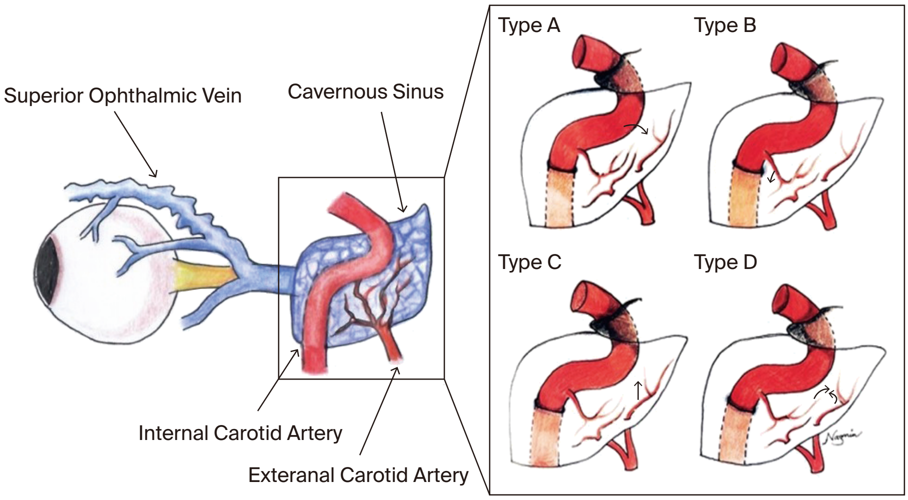

Figure 2. Types of carotid cavernous fistulas. Source: Figure by authors.

4. Pathophysiology

The underpinning pathophysiology (Greenberg 2010; Korkmazer et al. 2013; Ertl et al. 2019) for the clinical consequences of all caroticocavernous fistulas (CCFs) is characterized by elevated intracavernous venous sinus pressure. This is also likely to lead to retrograde venous drainage to the eye by the superior ophthalmic vein. Because of the interconnections between the two cavernous sinuses, contralateral eye involvement is common. Occasionally, due to occlusion of the superior ophthalmic vein by anterior intracavernous thrombosis, the clinical presentation may only occur contralateral to the fistula.

5. Natural History of CCFs

The natural history for the eye and vision is poor in extreme cases. Threats to vision can arise from a secondary glaucoma and extreme exophthalmos with consequent corneal damage. In the case of indirect CCFs, the clinical manifestations may be due to the combination of cavernous sinus thrombosis and a DAVF. In such cases, an extremely small DAVF may be responsible for extreme clinical manifestation. In the case of a direct CCF, retrograde flow in the ophthalmic artery may occur, contributing to retinal ischemia that, when combined with the high venous pressure, may lead to immediate permanent loss of vision.

Problems other than those with the eye can occur. Retrograde cortical venous drainage may be present (middle cerebral veins or pontine venous tributaries to the inferior petrosal sinus). When present, the considerations discussed relating to DAVFs need to be taken into account (Greenberg 2010; Macdonald 2008).

6. Etiology

Head or orbital injury;

Rupture of a cavernous ICA aneurysm;

ICA dissection (Greenberg 2010; Korkmazer et al. 2013; Ertl et al. 2019).

7. Clinical Presentation

7.1. Symptoms

Headache
Impaired vision
Double vision
Tinnitus (pulsatile)

7.2. Signs

Proptosis
Chemosis
Orbital bruit
Cranial nerve palsy
Corkscrew vessels of the conjunctiva
Increased intraocular pressure
Ophthalmoplegia
Ptosis
Venous pulsations
Heme in Schlemm's canal on gonioscopy (Greenberg 2010; Korkmazer et al. 2013; Ertl et al. 2019; Macdonald 2008; Kalangu et al. 2009; Bennett et al. n.d.).

8. Radiological Assessment

Besides a CT scan with a CTA (Figure 3) and MRI of the brain with an MRA (Figure 4), the definitive investigation method is a DSA of the brain to identify the feeding artery, draining vein and location of the AV fistula.

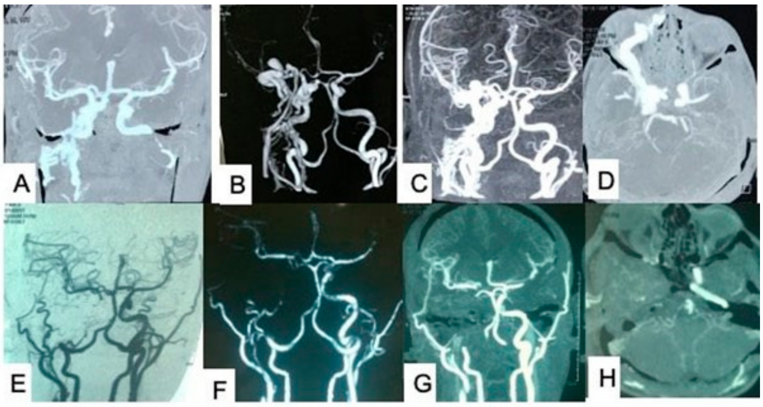

Figure 3. (**A**–**D**) Preoperative CTA showing a right-sided direct CCF. (**E**–**H**) CTA on the first POD after an STA–MCA bypass and ICA trapping. Source: Figure by authors.

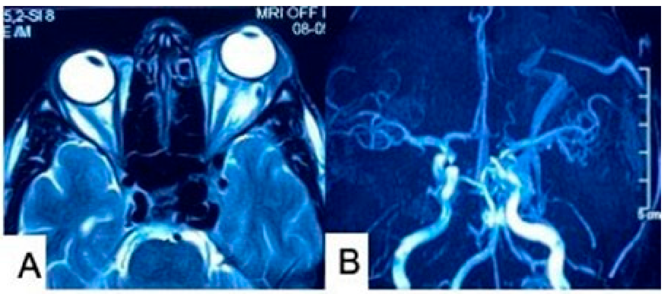

Figure 4. (**A**) MRI of the brain with an orbit showing the proptosis of the left eyeball, along with some irregularities in the cavernous sinus (left); (**B**) MRA showing the same constriction in the cavernous carotid artery, along with left-sided cavernous sinus dilatation and cortical venous drainage. Source: Figure by authors.

Proposed venous drainage-based classification system for CCFs;

Type	Venous drainage
I	Only posterior/inferior drainage
II	Posterior/inferior, as well as anterior drainage
III	Only anterior drainage
IV	Retrograde drainage into cortical veins ± other routes of venous drainage
V	High-flow direct shunt between a cavernous ICA and CS (Barrow type A) ± multiple routes of venous drainage (Kalangu et al. 2009).

9. Treatment of CCFs

Some traumatic CCFs may undergo spontaneous closure. There are various treatments for a CCF (Greenberg 2010; Cruz 1998; Korkmazer et al. 2013; Ertl et al. 2019; Macdonald 2008; Kalangu et al. 2009; Bennett et al. n.d.).

9.1. Carotid Compression

Carotid compression treatment may be successful in the closure of 17% of direct and 30% of dural CCFs.

9.2. Endovascular Management Is the Main Stay of Management

Approaches to carotid cavernous fistulas:

Transvenous routes via the following:

(a) Inferior petrosal sinus (IPS);
(b) Superior ophthalmic vein via the transfemoral route or direct surgical exposure;
(c) Facial vein via the transfemoral route;
(d) Transarterial route:

1. Direct CCFs are best managed with a detachable silicon balloon via an endo- arterial route;
2. Stent-assisted coil closure of a fistula may offer safe and effective management;
3. Onyx embolization by a transvenous route;
4. Microcoil embolization by a transvenous route;
5. Combination of Onyx and detachable coils through a transvenous route.

9.3. Surgical Trapping with or Without an STA–MCA Bypass

We commonly do surgical trapping of an ICA after thorough evaluation of the CCF by a DSA of cerebral vessels, cross-circulation study and balloon test occlusion (Figure 5).

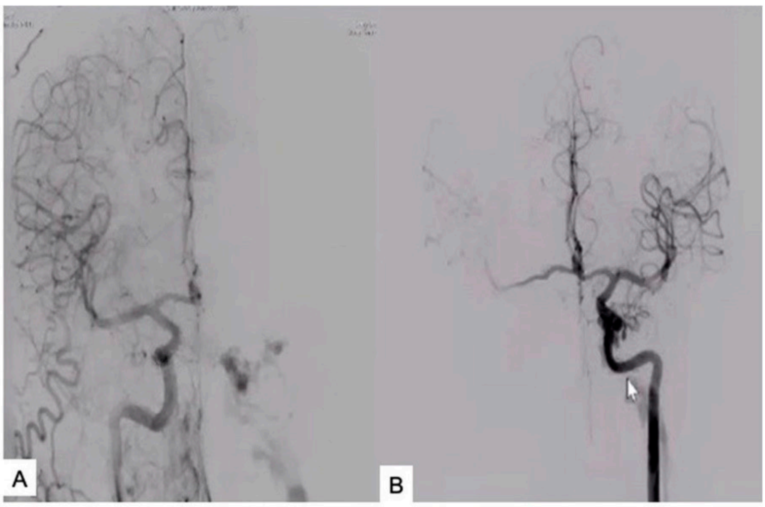

Figure 5. (**A**,**B**) Left and right cerebral ICA DSA, respectively, with a contralateral carotid occlusion in patients with a CCF. Source: Figure by authors.

We perform surgical trapping of the ICA by exposure and ligation at the high neck or Glasscock triangle in the middle fossa base, and ligation of a supraclinoidal ICA proximal to ophthalmic artery after anterior clinoidectomy. Some surgeons also use CS packing with trapping of the ICA.

[Balloon test occlusion: Before going to the occlusion or entrapment of an ICA, we need to evaluate the patency of the circle of Willis by performing a balloon test occlusion for half an hour.]

9.4. Gama-Knife Radiosurgery

In patients who fail or are unable to have an endovascular intervention, radiosurgical therapy of indirect CCFs has been advocated as a viable, non-invasive adjunct or primary treatment (Cruz 1998).

Author Contributions: Conceptualization, methodology, validation, formal analysis, investigation, resources, data curation, F.H.C., S.A. and N.A.; writing—original draft preparation, F.H.C. and S.A.; writing—review and editing, F.H.C.; visualization, supervision, N.A. and M.R.H. All authors have read and agreed to the published version of the manuscript.

Funding: This research received no external funding.

Conflicts of Interest: The authors declare no conflicts of interest.

References

Barrow, Daniel L., Robert H. Spector, Ira F. Braun, Jeffrey A. Landman, Suzie C. Tindall, and George T. Tindall. 1985. Classification and treatment of spontaneous carotid-cavernous sinus fistulas. *Journal of Neurosurgery* 62: 248–56. [CrossRef] [PubMed]

Bennett, Jeffrey L., Mithra O. Gonzalez, and Vikram Durairaj. n.d. Carotid-Cavernous Fistulas. Available online: https://www.uptodate.com/contents/carotid-cavernous-fistulas (accessed on 31 January 2021).

Chowdhury, Forhad, Mohammod Haque, Khandkar Kawsar, Shamim Ara, Quazi Mohammod, Mainul Sarker, and Atul Goel. 2012. Transcranial Microsurgical and Endoscopic Endonasal Cavernous Sinus (CS) Anatomy: A Cadaveric Study. *Journal of Neurological Surgery* A73: 296–306. [CrossRef] [PubMed]

Cruz, Julio. 1998. *Neurologic and Neurosurgical Emergencies*. Philadelphia: WB Saunders Company.

Ertl, Lorenz, Hartmut Brückmann, Maximilian Patzig, and Gunther Fesl. 2019. Endovascular therapy of direct dural carotid cavernous fistulas—A therapy assessment study including long-term follow-up patient interviews. *PLoS ONE* 14: e0223488. [CrossRef] [PubMed]

Greenberg, Mark S. 2010. *Handbook of Neurosurgery*. New York: Thieme.

Kalangu, Kazadi, Yoko Kato, Gilbert Dechambenoit, and Hirotoshi Sano. 2009. *Essential Practice of Neurosurgery*. Nagoya: Access Publishing Co., Ltd.

Korkmazer, Bora, Burak Kocak, Ercan Tureci, Civan Islak, Naci Kocer, and Osman Kizilkilic. 2013. Endovascular treatment of carotid cavernous sinus fistula: A systematic review. *World Journal of Radiology* 5: 143–55. [CrossRef] [PubMed]

Macdonald, R. Loch. 2008. *Neurosurgical Operative Atlas: Vascular Neurosurgery*. New York: Thieme.

Tang, Yen Zhi, Thomas Booth, Michael Steward, T. Solbach, and Thomas Wilhelm. 2010. The imaging of conditions affecting the cavernous sinus. *Clinical Radiology* 65: 937–45. [CrossRef] [PubMed]