Dural Arterio-venous fistulas: Abnormal arterio-venous shunts within the dura matter. 10-15% of all AVM...).

- **Aetiology** is enigmatic. The majority are **acquired idiopathic**. Cases with a documented history of trauma, surgery, venous thrombosis are not uncommon. Syndromes associated with vascular fragility such as fibromuscular dysplasia, Ehler-Danlos syndrome and NF1 have been associated with dural AVF.
- Primitive arterio-venous communications within the dura are known to involute as the dura matures. Microscopic remnants of these communications are found in adult dura. Venous sinus thrombosis, infection, trauma and possibly hormonal and haemostatic factors may cause reopening of these channels and the formation of a fistula. This fistula may thrombose and involute spontaneously or it may recruit additional arterial supply. This will lead to **venous hypertension.** Venous sinus obstruction leads to **intracranial hypertension** and reversal of the blood flow to leptomeningeal channels which causes venous dilatation, varicosities and venous aneurysms which predisposes to intracranial haemorrhage. **Few cases are congenital**.
- **Presentation**: see below for symptoms depending on location

1. Haemorrhage: The key factor in terms of natural history is the presence or absence of leptomeningeal venous drainage. **The annual risk of haemorrhage is 1.6% in the presence of leptomeningeal venous drainage.**
2. Non haemorrhagic symptoms may secondary to venous hypertension (hydrocephalus and pseudotumor cerebri).
3. In case of cavernous AVF (orbital and ocular congestion, ophthalmoparesis, glaucoma)
4. Pulsatile tinnitus

- Investigations:

1. CT-haemorrhage (bone windows can show grooving of the produced by dilated veins and arteries)
2. MRI-flow voids. MRV-can show venous sinus narrowing and or thrombosis
3. cerebral angiogram including the external carotids is the most important test, in particular the **venous phase** looking for the **direction of venous flow**, narrowing or obstruction of venous sinuses and the presence or absence of cortical leptomeningeal venous drainage. In many cases subclavian branches including thyrocervical and costocervical arteries should be included.

- **Borden classification** takes into account the pattern of venous drainage (Type1-no venous obstruction and drainage is into the sinus, Type2-has some degree of venous obstruction and the drainage is both in the sinus and into cortical veins, Type 3-complete obstruction of the sinus and drainage is only into the cortical vein. Both type 2 and 3 follow an aggressive course.
- Treatment is indicated only for fistulas with intolerable progressive symptoms and those **deemed to be at high risk of bleeding and neurological deficit** (haemorrhge, venous infarct, visual loss and neurological deficit). Partial treatment offers no protection against recurrent haemorrhge. Asymptomatic lesions and those with low risk are observed. Patients should report to the physician any change in symptoms (disappearance of bruit may indicate thrombosis of the fistula and cure or development of leptomeningeal venous drainage and increased risk).
- Treatment options:

1. Compression of OA or STA for 30 min every day occassionally results in thrombosis of ST or sagittal DAVF -25%
2. Endovascular embolisation
    A. **Tranvenous** embolisation through transfemoral or direct approach through burr hole is the treatment of choice for most DAVF and carries high cure rate and minimal complications.
    B. **Transarteria**l embolisation: **cure rate <50%.**
3. Surgical excision or disconnecting the fistula is occassionally indicated if the above methods fail

- Symptoms and treatment depend on the type and location of the fistula:

1. **Transverse/ sigmoid**: The most common type (**50-63%)** - the most common symptoms are **pulsatile tinnitus**, headaches. The venous drainage is into the sinus in the vast majority of cases and not into leptomeningeal veins, hence the clinical course is **benign**. Rarely they produce venous HTN and present with papilledema and visual obscuration (10%). Intracranial haemorrhge is even rarer. Feeding arteries are branches of occipital, APA, STA, posterior auricular, middle meningeal and branches of ICA (meningohypophyseal trunk, tentorial branch, and inferolateral trunk) and posterior dural branch and muscular branch of VA and occassionally pial branches. Venous drainage is either into sigmoid-transverse sinus, cortical veins or both depending on the type .Asymptomatic lesions are observed. Symptomatic lesions and those with leptomeningeal venous drainage require treatment directed at disconnecting the abnormal leptomeningeal venous channels. **Transvenous embolisation** using GDC is the treatment of choice. If the sinus is thrombosed it can be excised with the fistula.
2. **Cavernous**: orbital congestion, ophthalmoparesis, headaches. Asymptomatic lesions are observed. Symptomatic lesions are treated with stereotactic radiosurgery + postradiation embolisation (transarterial or tranvenous "superior orbital vein".
3. **Anterior fossa**: **high risk** of intracranial haemorrhage. These are high risk dural AVF and almost always have leptomeningeal venous drainage. Venous aneurysms (varices) are common. Feeding arteries are the ethmoidal branches of the ophthalmic artery. Treatment is surgery through subfrontal approach. The goal is to **disconnect the leptomeningeal arterialised venous channels**.
4. **Petro-tentorial**: **high risk** of haemorrhage from leptomeningeal veins. Arterial feeders are from the tentorial branches of ICA, ascending pharyngeal artery and vertebrobasilar system. Treatment is surgical disconnection of the leptomeningeal draining veins via **subtemporal** or other **base of skull approaches**
5. Sagittal sinus: 7% of all DAVF. **50% have leptomeningeal drainage** and can have aggressive course. Symptoms depend on the pattern of venous drainage. If the drainage is solely in the sinus, patients present with headaches, bruit and occassionally venous hypertension (hydrocephalus and pseudotumor cerebri). If the venous drainage is shared by cortical veins, there is high incidence of ICH and neurological deficit. Feeders are from STA, MMA, OA, occassonally falcine branch of ophthalmic artery, MHT through Bernasconi-Cassinari artery) Asymptomatic lesions are observed. Symptomatic lesions are treated by disconnecting the leptomeningeal vessels either by embolisation (Transvenous or Transarterial) or surgically.

Vein of Galen malformations:

- The vein of Galen is a short, centrally located vein in the pineal region under the splenium of corpus callosum. It is formed by the confluence of two internal cerebral veins and two basal veins of Rosenthal. Embryologically it is derived from the posterior part of the median prosencephalic vein of Markowski which joins the internal cerebral veins and basal veins at 3 week of gestation.
- Vein of Galen malformations are **high flow shunts** with three modes of presentation depending on the age 1. **Neonates** present with **high output cardiac failure** (machinery murmur, tachycardia, tachypnoea and metabolic acidosis). It carries a high mortality 50-80% 2. **Young children** present with **hydrocephalus** and increasing head circumference. Dilated periorbital 3. Old children and adults present with SAH and learning difficulties.
- Pathologically this is a high flow arteriovenous shunt similar to dural AVF.. Yasargil described four types depending on the nature of feeding arteries:

1. Type 1: arterial feeders from pericallosal and posterior cerebral arteries
2. Type 2: fed by thalamoperforating and posterior cerebral arteries.
3. Type 3: Mixed pattern of feeders 1+2
4. Type 4: parenchymal malformations behave as true AVM

- Diagnosis 1. Neonatal ultrasound 2. CT head (calcifications from long standing ischemia 3.MRI/MRA- hydrocephalus, parenchymal injury from ischemia, vein of Galen aneurysms 4. Cerebral angiogram is the gold standard for defining the complex anatomy and therapeutic planning.
- **Treatment** of these lesions has evolved towards **endovascular embolisation** via transvenous or transarterial approaches. **Transtorcular transvenous embolisation of the fistula is a popular method.** Surgery, particularly in the neonates with high output cardiac failure is associated with high mortality and morbidity.
- The **role of surgery** is to manage **hydrocephalu**s, to provide access **(Transtorcular approach**), and to **manage Type 4 malformations which should be approached as AVM.**
- The two major complications of endovascular treatment are 1. Perforation of the venous aneurysm by the guide wire-SAH 2. Complete occlusion of the venous outlet leading to venous hypertension and haemorrhage. Mortality is high in the neonates presenting with high output cardiac failure 50-80%.