"Perinatal imaging of congenital intracranial vascular malformations".

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Learning objectives

Perinatal imaging of congenital intracranial vascular malformations (CIVM) comprise the use of the different available imaging modalities to reach an adequate diagnosis of these rare malformations in the prenatal and postnatal period of life.

Ultrasound (US) and MRI are normally used in the prenatal assessment of fetuses with CIVM. In the neonatal period, until one month of age, it will be necessary a chest plain film, brain and cardiac US, a cranial Angio computed tomography (ACT) and/ or MR imaging (AMRI). The role of conventional arteriography as a diagnostic and therapeutical procedure will be essential in some neonates with CIVM.

Accurate interpretation of all these examinations requires an understanding of the anatomy, physiology, distribution, and flow patterns of the fetal and neonatal brain vasculature.

The learning objectives are:

- Illustrate the two more common types of these rare malformations in the perinatal period.
- To indicate the difference of CIVM in the fetus and neonate as compared to the approach of CVIM in adults and older infants and children.
Background

CIVM are very rarely detected in the perinatal period. However, the improvement in perinatal imaging diagnosis, better neonatal intensif care and the possibility of endovascular treatment in tertiary referral care centers have made necessary to radiologists to deal with these complicated cases in the fetal and neonatal period.

CIVM that appear in perinatal period of life comprise non proliferative arteriovenous (AV) lesions or shunts in the brain. They can be located in the:

- epidural/subdural space, like the dural sinus malformation or DSM.
- subarachnoid space, like the vein of Galen malformation or VGAM.
- subpial space, the origin of the pial arteriovenous malformation or PAVM.

Dural sinus malformation (DSM)

Congenital DSM are AV shunts usually seen as a giant dural lake with very slow flow communication with the other sinuses. They can be located:

- in the mid-line, involving the superior sagittal sinus (SSS) and the torcular, a location implying very poor outcome.

- lateralised to the tranverse sinus, normally with better outcome.

In almost all cases there is either thrombosis or a hypogenetic jugular bulb.

In the neonatal period they may present as a mild cardiac failure, coagulation disorders or increased intracranial pressure (seizures, etc). Poor prognostic indicators include mid-line position, large size of pouch and postnatal thrombosis of the pouch.

DSM can suffer spontaneous thrombosis prenatally and if it is rapidly and extensive coagulation factor consumption can be a consequence and worsen the prognosis.

Accurate prenatal diagnosis of DSM is essential to providing parents with information upon which to base decisions. The current prognosis of DSM postnatally is generally unfavorable because spontaneous thrombosis can compromise the venous drainage of the brain with subsequent venous infarction and intraparenchymal hemorrhage.
Nonetheless, good outcomes have been referred in some cases in the literature, with progressive involution of the DSM secondary to the spontaneous thrombosis and posterior adequate neurological development.

Vein of Galen malformation (VGAM)

VGAM is the most frequent of the CVIM. It consists in an abnormal AV fistula, connecting deep choroidal arteries and the median prosencephalic vein of Markowski, resulting in a dilated venous pouch or varyx in the vein of Galen region.

Based on the angioarchitecture of the VGAM, two types have been described: choroidal, with multiple feeders from pericallosal, choroidal and perforating arteries, and mural, receiving a single or few feeders from posterior arteries.

In fetal period the presence of fetal cardiomegaly, hydrops, severe ventriculomegaly and ischemic brain lesions usually detected by MR indicate poor outcome.

The most severe forms of VGAM produce early brain ischemia (cerebral effect of the hemodynamic steal). Less severe forms are expressed by perinatal high output congestive heart failure (CHF). Liver and renal failure can present secondary to CHF.

In more benign forms the patients are normal at birth, but they develop progressive hydrocephalus in the first postnatal months.

The severity of the clinical picture depends on the importance of the arteriovenous shunt. In big malformations lack of treatment rapidly leads to multiorgan failure and a cerebral melting process within days or weeks.

Pial arteriovenous malformation (PAVM)

The perinatal diagnosis of PVAM is extremely rare and only a few cases have been reported in the literature.
Findings and procedure details

VGAM

PRENATAL IMAGING FINDINGS

1. Prenatal brain US

The detection of a large fetal intracranial mass as a pseudocystic, non-echogenic, tubular or spheric-shaped mid-line lesion represents a VGAM or a DSM (Fig.1). PAVM is very rarely detected prenatally.

US can identify the VGAM and associated complications as ventriculomegaly.

The use of Doppler imaging (Fig. 2 & 3) is mandatory in these fetuses not only to make the right diagnosis but to establish fetal brain vascular changes and identify collaterals and arterial feeders indicating the type and complexity of the malformation.

Doppler imaging with simultaneous color and spectral Doppler interrogation of the major cerebral arteries and veins render valuable functional and hemodynamic information like high fistulous flow in the VGAM or vasodilation of a major cerebral artery (Fig. 3).

2. Fetal MRI

Prenatal MRI should be considered in the end of the second or in the third trimester to improve brain damage assessment and therefore allow adequate parental counseling.

MR better shows associated findings such as hydrocephalus and brain atrophy, and can differentiate VGAM from a cerebral arteriovenous malformation draining into the vein of Galen.

The use of ultrafast T2-weighted MR sequences like single-shot fast spin echo (SSFSE) have made possible to detect as flow voids the dilated and abnormal
brain vessels in VGAM (Fig. 4) and also early ischemic lesions as abnormal hyperintensities in surrounding brain parenchyma (Fig. 5).

NEONATAL IMAGING FINDINGS


The first postnatal imaging technique in these patients should be a chest X-ray and an echocardiography in order to evaluate the secondary cardiac failure than can often be associated to prenatally detected VGAM (Fig. 6).

2. Neonatal brain US

The angioarchitecture of the VGAM can be studied with depiction of the dilated feeding vessels as well as the dilated vein of Galen by Doppler US (Fig. 7). In addition, the degree of brain edema, hydrocephalus, and possible thrombosis can be studied.

Besides, Doppler US may be used to evaluate the success of neurointerventional treatment.

3. Cranial Angio CT or MRI.

Although Gadolinium-enhanced cranial MR venography is preferred to CT in newborns, in ill patients with complicated VGAM (CHF, mechanical ventilation in the NICU, etc) cranial angio CT is usually performed.

Maximum intensity projections or MIP (Fig. 8) or three-dimensional (3D) reconstructions are especially helpful for better understanding the angioarchitecture of vascular lesions as well as their relation to the intracranial vasculature. Highend post-processing programs allow arterial (CT angiography) and venous (CT venography) detailed mapping pre endovascular treatment.

MRI is the best technique in the postnatal period to study VGAM and the possible complications, like brain parenchyma ischaemic oedema, with the major benefit of absence of irradiation or the use of iodine contrast agents.

4. Angiography.
Although it is been demonstrated that transcatheter embolization of perinatal detected VGAMs have better prognosis when performed at least after 5-6 months of age, sometimes the clinical situation of the patients make necessary endovascular treatment in the first three months of age (Fig.9).

The technical description of the angiography and endovascular treatment of VGAM is beyond the scope of this educational exhibit.

DSM

PRENATAL IMAGING FINDINGS

1. Prenatal US.

US can detect a hypoechoic heterogeneous lesion, located in the posterior fossa, displacing cerebellar vermis towards the fourth ventricule. It has to be differentiated from an arachnoid cyst, as a low flow at Doppler can be misinterpreted as bleeding inside a retrocerebellar cyst.

2. Fetal MRI.

Used as a second step examination to assess the diagnosis and, as far as possible, the prognosis. The characteristic dural location of the mass centered on the torcular is exquisitely depicted by MRI (Fig.10).

The thrombosis, seen as an heterogeneous pattern on the T1-weighted sequences including hyperintense areas, and furthermore, the progressive changes observed during the follow-up constitute interesting diagnostic criteria.

Other MR sequences as Diffusion weighted imaging or gradient echo T2* are useful in these fetuses looking for brain ischaemic or hemorrhagic complications.

NEONATAL IMAGING FINDINGS

1. Brain Doppler US.
US should be the first step in postnatal imaging in DSM detected prenatally. A posterior fossa hyperechoic and heterogenous lesion is often detected although another technique will be needed to rule out other complications.

2. Cranial Angio CT or MRI.

When DSM suffers thrombosis a heterogeneous extra-axial mass with variable T1 signal intensity (Fig.11) and T2 signal intensity components can be seen. The areas of high T1 signal intensity are compatible with areas of thrombus.

This mass are ususllay centered on the torcular region and can compress the cerebellum anterior or inferiorly. Ventriculomegaly of the third and lateral ventricles can also be present.

Haemorraghic or more rarely ischaemic events can appear in the neonatal period and must not be overlooked.

3. Angiography.

The use of transcatheter endovascular treatment have been described but normally in older infants.
Fig. 1: 35 week-old fetus brain US. A posterior mid-line hypoechoic tubular lesion is seen (white arrow). Surrounding smaller tubular structures can also be depicted.

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**Fig. 2:** Colour Doppler imaging of the same fetus than Fig.1 demonstrates the vascular nature of the brain lesion (white arrow).

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**Fig. 3:** Spectral Doppler of the middle cerebral artery of the fetus indicates high diastolic flow secondary to dilation of major brain vessels.

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**Fig. 4:** Coronal T2-FSE weighted image of a 36 week-old fetus. A big aneurysmatic sac can be seen in the mid-line representing the VGAM (white star). Surrounding multiple arterial (choroidal) vessels feeding the malformation (red arrow). Right parietal ischaemic changes can already be seen.

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Fig. 5: Sagittal T2 FSE weighted image of the same fetus than fig.4. Pericallosal dilated arteries (white arrow) and a big dilated VGAM are nicely shown.

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Fig. 6: Chest plain film of a term newborn prenatally diagnosed of VGAM. We can see remarkable cardiomegaly and perihilar vascular increased marks representing pulmonary oedema.

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Fig. 7: Colour Doppler US of a neonate with prenatal findings of VGAM. High flow is seen in the aneurysmatic malformation (white arrow). Dilated pericallosal arteries running towards the VGAM are also seen.

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**Fig. 8:** Postnatal contrast-enhanced brain CT (MIP reconstruction of the sagittal plane) can show the important dilation of the vascular malformation including the torcular area (black arrows). Multiple collateral feeding arteries from the anterior and middle circulation are also shown (white arrow).

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Fig. 9: Three week-old newborn diagnosed of VGAM. Brain angiography after inner carotid catheterization. Anterior and choroidal arterial branches feeding the malformation (red arrows).

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**Fig. 10:** Transverse fetal FIESTA MR image showing a posterior dilated venous sinusal pouch, involving the torcular (white star).

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Fig. 11: Postnatal T1-weighted MRI in the sagittal plane shows the hyperintensity and heterogenous thrombosis in a DSM (white star).

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Conclusion

CIVM are rare in perinatal period but their prenatal detection is becoming more frequent due to improved fetal imaging, including the supplementary role of fetal MRI.

Early diagnosis is important in the management of VGAM and DSM regarding the parental counseling during the pregnancy and postnatal assessment.

The use of multimodality imaging is mandatory in the neonatal period to evaluate brain and systemic complications of these vascular malformations and state the feasibility of endovascular treatment.
References