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Mini Review

The Importance of Ultrasound in Diagnosing Congenital Vascular Anomalies in the Fetal Brain?

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Abstract

Congenital Vascular Anomalies are very rare and occur in only roughly one percent of births. Since it is so rare, doctors have trouble diagnosing these cases because of uncertainty with the disease. The role of ultrasound can play a major role in detection and diagnosis of any congenital vascular anomaly developing in the fetal brain, leading to proper and quick possible treatment. Vascular anomalies represent the most common cause of pediatric soft-tissue masses [1]. These vascular anomalies include vascular malformations, vascular tumors and congenital vascular defects. The incidence of Arteriovenous Malformations estimated occurrence rate is one in every 100,000 [2]. An estimated two-thirds of AVM's occur before the age of 40 [2]. Every year, roughly four out of 100 people with an AVM will experience a hemorrhage and each hemorrhage poses a 20 percent risk of death or stroke, 30 percent neurological morbidity, and 10 percent mortality [2].

Keywords: *Ultrasound; Congenital Vascular Anomalies; Fetal Brain*

Introduction

Vascular malformations

Vascular malformations of the brain is a term for six different conditions in which blood vessels in the brain are affected. These malformations are classified into several types of malformations in which the severity, symptoms and causes may vary. The types of Vascular Malformations of the Fetal Brain include: 1) Arteriovenous Malformations (AVM), abnormal arteries and veins; 2) Cavernous Malformations, enlarged blood-filled spaces; 3) Venous Angiomas (malformations), abnormal veins; 4) Telangiectasias, enlarged capillary-sized vessels; 5) Vein of Galen Malformations; and 6) Mixed Malformations [3]. Other than vascular anomalies the following fetal congenital anomalies can also be evaluated by 2D grayscale Ultrasound and they are: 1) Anencephaly, 2) Cephalocele, 3) Hydrocephalus, 4) Hydrancephaly, 5) Spina Bifida, 6) Meningocele, 7) Myelomeningocele, 8) Arnold- chiari- malformation, 9) Dandiwalker malformation, 10) Cervical Teratoma, 11) Cystic hygroma, 12) Holoprosencephaly, 13) Hypophosphatasia, 14) Facial anomalies, 15) Microcephaly, 16) Macrocephaly, etc.

Discussion

Arteriovenous malformations

Arteriovenous malformations are abnormal masses of blood vessels that cause multiple irregular connections between the arteries and veins. In an AVM, arteries connect directly to veins without a capillary bed in between. This creates a problem called a high-pressure shunt or fistula [4]. In this situation veins are not able to handle the pressure of the blood coming directly from the arteries, causing the veins to enlarge and stretch beyond their capacity. As a result, the weakened blood vessel can rupture and bleed into the brain, which can

cause stroke, brain damage, and even death. Other neurological symptoms that may also occur include: headache, weakness, seizures, pain, problems with speech, visions or movement. These malformations usually occur in the spinal cord and in any part of the brain or its surface, but can occur anywhere in the body [5]. Approximately 50 percent of patients present initially with a bleed, while other times patients may not experience any symptoms [2]. An estimated 12 percent of people with an AVM will experience symptoms [2]. Symptoms associated with AVM are: Seizures, Muscle Weakness or Paralysis, Loss of Coordination, Dizziness, Headaches, Visual Disturbance, Language Problems, Mental Confusion, Hallucinations, and/or Dementia.

An AVM may affect the following functions of the brain: the frontal lobe functions to process motor movements, regulates personality and articulation of speech; the parietal lobe functions to process sensory information, such as sensation of pain, temperature, light touch and more; the temporal lobe functions to process things related to hearing, memory, learning and receptive speech; and the occipital lobe functions to process things related to vision [6]. They can also effect the back part of the brain (cerebellum), the brainstem or the ventricles (deep spaces within the brain that produce and circulate cerebrospinal fluid). AVM's usually start to develop while the baby is still in the uterus. It is known to result from an error in embryonic or fetal development [3]. Although AVM's are congenital they do not necessarily run in families and they are not inherited from ones parents. A brain AVM contains abnormal or weakened blood vessels that direct blood from normal brain tissue. These blood vessels dilate over time and can eventually rupture and cause bleeding into the brain. More than 50 percent of patients may present with an intracranial hemorrhage [6]. Among AVM patients, 20 to 25 percent may experience focal or generalized seizures. Patients may experience headaches due to increased blood flow around the anomaly and 15 percent may have trouble with vision, speech and/or movement [6].

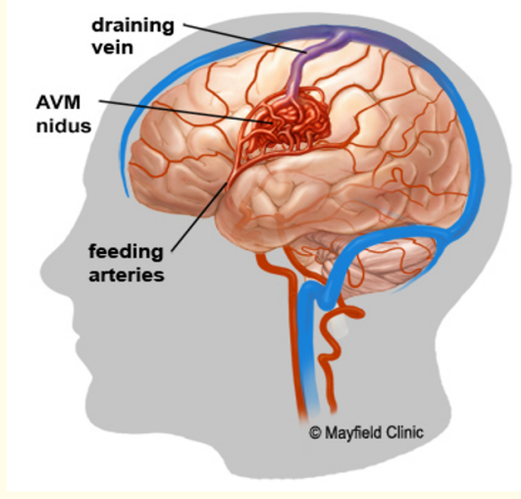

Figure 1: *Shows an AVM is a tangled bundle of blood vessels where arteries connect directly to veins with no capillary bed between.*

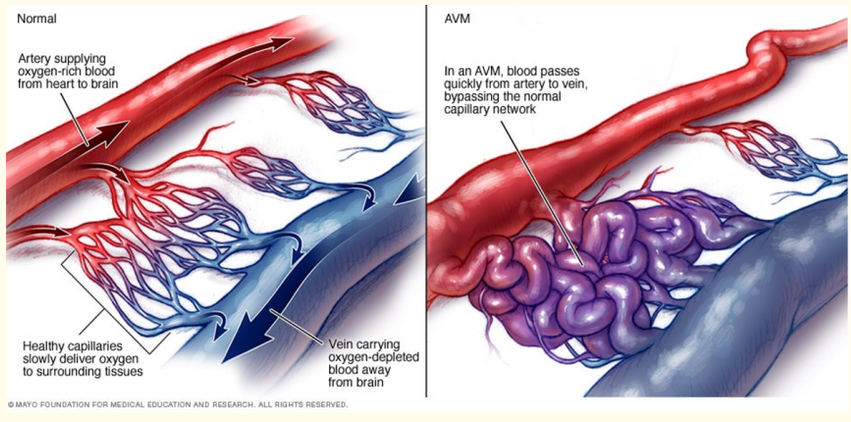

Figure 2: *Shows a normal blood vessels compared to an AVM.*

Cavernous malformation

Cavernoma (Cavernous malformation) is an abnormal cluster of enlarged capillaries with no significant feeding arteries or veins; abnormal formation of the blood vessels; low pressure [4]. These clusters of capillaries grow with extremely thin walls, increasing the chances of the walls to bleeding. When these malformations grow in the brain they increase chances of headaches and/or seizures. Very often these malformations do not cause any symptoms and no detection is made. In some cases, the lesions can burst and bleed into the brain, causing neurological problems, stroke and even death. These malformations account for approximately 5 - 15% of all vascular abnormalities in the central nervous system [7]. Cavernous Malformations are extremely rare in the fetal and neonatal period.

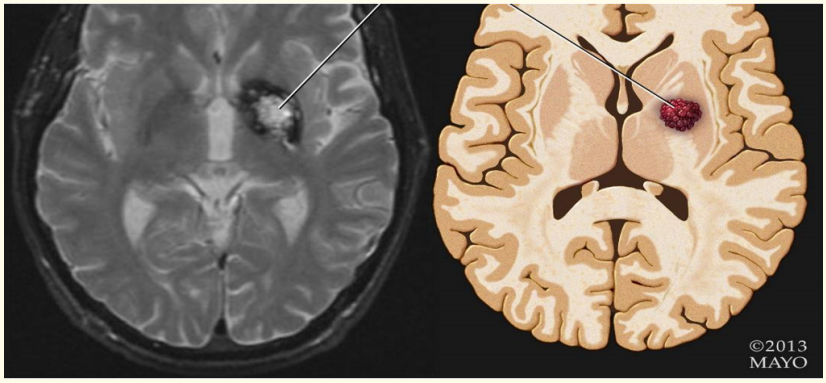

Figure 3: *Shows an MRI image of a cavernous malformation.*

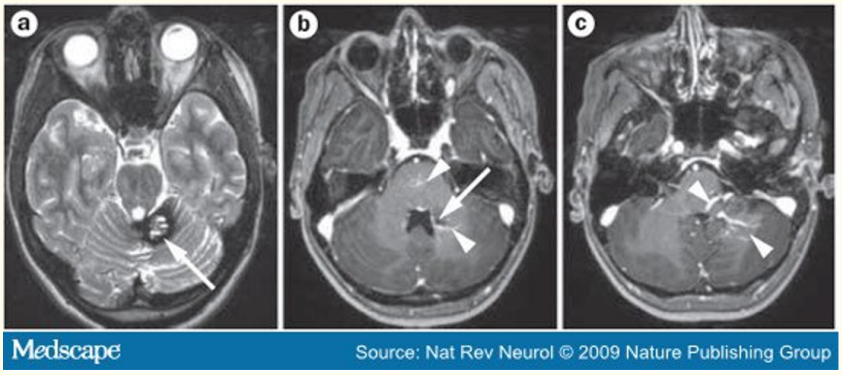

Figure 4: *Shows venous anomalies associated with cavernous malformations [8].*

Venous malformations

Venous malformations (Angiomas) are abnormal clusters of enlarged veins. In the brain the network of veins work to drain the organ of deoxygenated blood. Most venous malformations do not show any symptoms and do not give rise to many complications. However, when they are combined with other lesions such as cavernous malformations, they may cause headaches, bleeding in the brain, numbness, weakness in the arms or legs, vision problems, speech and memory problems, seizures, and ischemic stroke [9]. If treatment is found to be necessary it can be limited to just monitoring the malformation by diagnostic ultrasound screening.

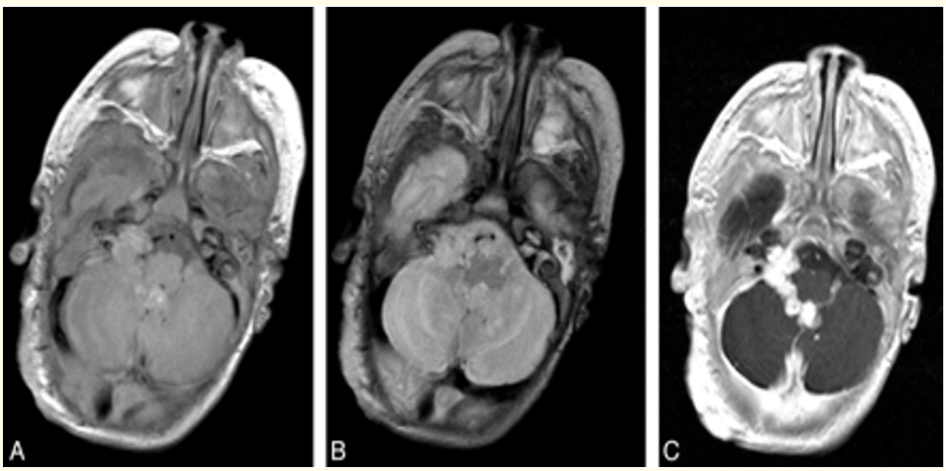

Figure 5: *Shows an MRI of an infant brain with a hemangioma [10].*

Capillary telangiectasia

Capillary telangiectasia, abnormal capillaries with enlarged areas (similar to cavernoma); very low pressure, rarely bleed and usually not treated. Group of abnormally swollen capillaries that are benign and rarely cause any symptoms. These are normally found in the pons, but are also said to be located in the medulla, caudate nucleus, cerebrum, cerebellar hemispheres, and in the spinal cord [11].

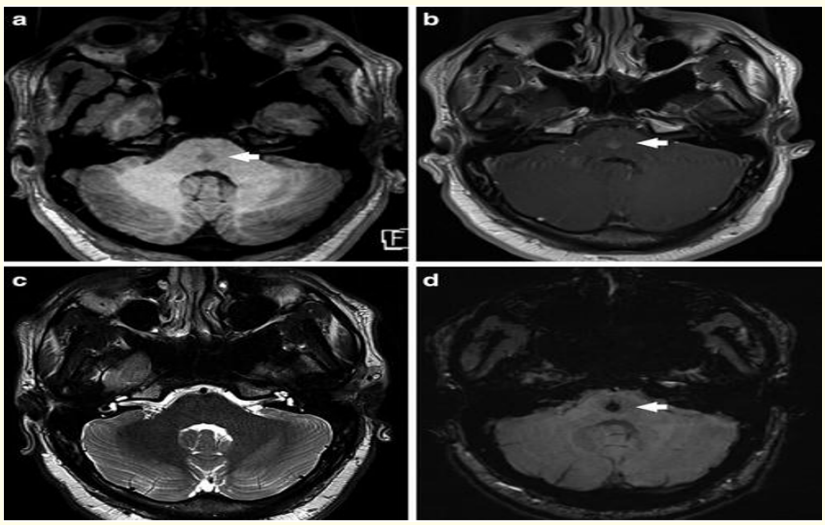

Figure 6: *Shows a capillary telangiectasia.*

Vein of galen malformations

Vein of galen malformations is an abnormal tangle of blood vessels that disrupt the normal flow of blood. In this abnormality the tiny capillaries that normally distribute oxygen-rich blood throughout the brain are missing. Since the capillaries are missing blood rushes into the Vein of Galen with too much force causing the heart to work vigorously eventually leading to potential heart failure [12]. This abnormal blood flow into the veins can also interfere with the normal balance between Cerebral Spinal Fluid (CSF) production and absorption, leading to hydrocephalus [12]. When a Vein of Galen Malformation is the most common cerebral arteriovenous malformation detected prenatally and accounts for 30% of all pediatric vascular malformations [13]. Diagnosis is often made prenatally in the third trimester

through the use of 2D and 3D ultrasound and magnetic resonance imaging (MRI) [13]. A transabdominal ultrasound assessment most often includes neurosonography and echocardiography of the fetus. These ultrasound tests show detailed grayscale, color and power doppler evaluation of the Vein of Galen Malformation.

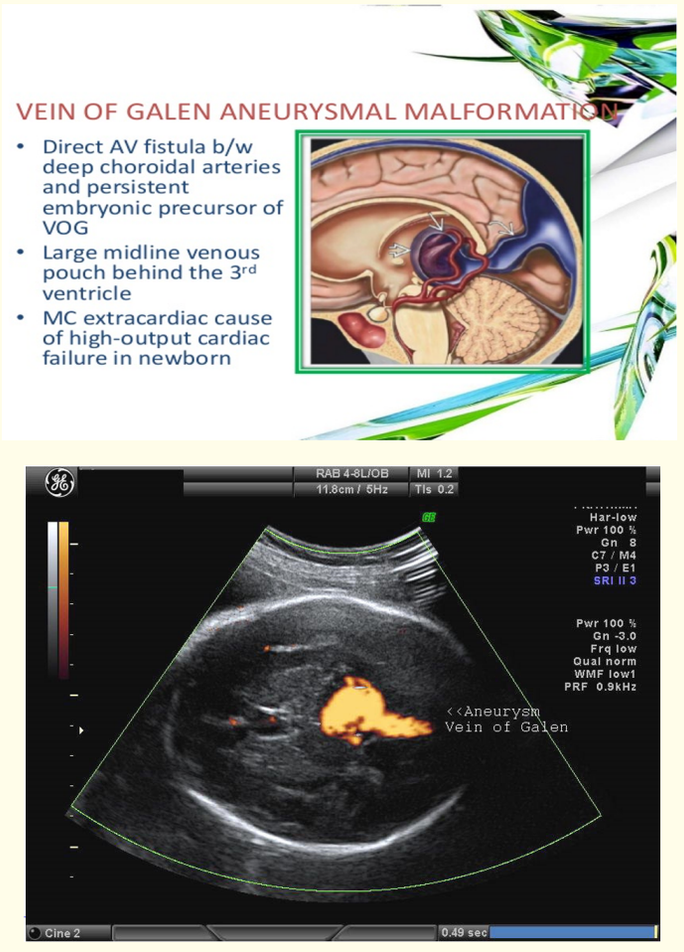

Figure 7: *Shows vein of galen aneurysm.*

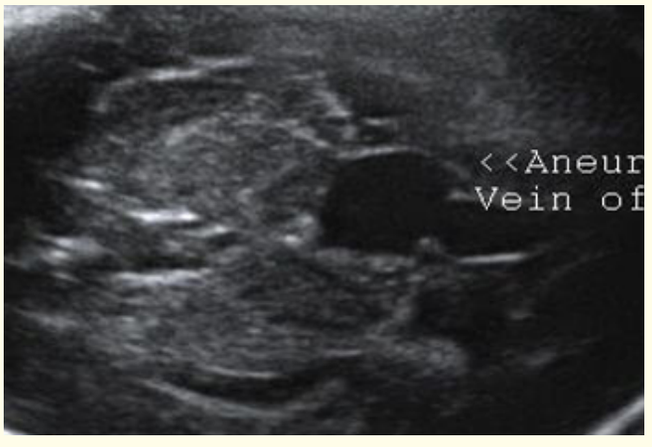

Figure 8: *Shows an intracranial venous aneurysm in fetus.*

Ultrasound consists of multiple tests and scanning techniques during pregnancy and postpartum. Screening for brain anomalies in the fetal head during pregnancy may consist of transabdominal and transvaginal sonography, along with Color and Power Doppler during the second and third trimesters and shortly after birth. During the neonatal period scanning the fetal head is accomplished by using a high-frequency phased array transducer (5 - 8 MHz) with a small footprint probe. High resolution images are obtained in preterm neonates by using probe frequency of 7.5 MHz. Most examinations are performed bedside with the neonate within the incubator. Neurosonography starts with gray-scale imaging performed via the anterior fontanelle in the coronal and sagittal planes. Color Doppler images may be obtained for vascular structures. Power Doppler imaging should be used to distinguish areas of hypo- or hyper- vascularity in suspected vascular anomalies or occlusions. Doppler examination makes diagnostic confirmation possible which increases treatment options.

Treatment possibilities include Open Brain Surgery which removes the abnormal connection, this surgery is done through an opening in the skull. Another method is Endovascular Embolization, a catheter is guided through a small cut in your groin. It then enters an artery and into small vessels that lead to the brain where the aneurysm is located [14]. Finally, a glue-like substance is injected into the abnormal vessels, this stops blood flow in the AVM and reduces chances of bleeding. A third method of treatment is Stereotactic Radiosurgery where radiation is aimed directly on the area of the AVM. This method shrinks the AVM and reduces bleeding [14]. It is helpful for small AVM's located deep in the brain where surgery is normally too difficult [15-21].

Conclusion

In summary, ultrasound provides an excellent diagnostic tool when fetal intracranial masses are recognized. It should be applied along with Doppler assessment of the mass to determine vascularization and to visualize proper blood flow. Over recent years, widespread use of ultrasound, computed tomography, and magnetic resonance imaging has lead to an increase in antenatal and postnatal detection of brain anomalies [13].

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