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Review Article

Role of Doppler Ultrasound in the Prenatal Diagnosis of Vein of Galen Malformations

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Abstract

Doppler ultrasonography is still the basis of routine ante-natal examinations. Identification of vein of Gallen malformation is usually prompted by cardiac anomalies. A small vascular lesion can be picked up by ultrasound when there is evidence of ventriculomegaly or echogenic defects in the ultrasound examination. Colour doppler studies are superior to routine ultrasound examinations . However there are certain drawbacks which need to be identified by the examiner.

Keywords: *Vein of Galen Malformations; Ultrasound Diagnosis; Antenatal*

Abbreviations

VG: Vein of Galen; VGM: Vein of Galen Malformations; AM: Arteriovenous Malformations

Introduction

Antenatal ultrasonic examination of pregnant women is a very commonly performed routine procedure. The diagnosis of vascular malformation by doppler ultrasonography is very rare. The most common lesion that is picked up by this ultrasonic method is the aneurysm of the vein of Galen (VGM). The diagnosis is usually made by the suspicion of cardiac failure and hydrocephalus.

There are many reports of diagnosis of VGM following ultrasonography. All these are single case reports. This is mainly because the diagnosis of the vascular malformation in the foetus is not very easy.

The following paper evaluates the whole of doppler ultrasonography in the diagnosis of vascular malformation and this paper also highlights some of the advantages of more recent diagnostic methodology such as colour doppler and magnetic resonance imaging of the foetal brain.

Literature Review

Lashaunda's [1-3] has tried to provide a theoretical concept of the formation of Arteriovenous Malformations (AVM). He refutes the congenital nature of the vascular formations. His concept is mainly based on the findings of Rendo Ossler Waver Disease. Unfortunately, the author does not consider the segmental pattern of the developing embryo which is essential in understanding any embryological anatomy. Therefore, his interpretation of the AVM and Vein of Galen formation at this stage remains purely speculative and is not supported by embryological or clinical evidence.

Sepulveda W, Platt, Fisk NM [4] published a paper on the pre-natal diagnosis of AVM using colour doppler ultrasonography. In this paper in addition to citing one of their own patients they collected 42 reported cases of VGM of the foetal brain diagnosed antenatally by ultrasonography. Of these 18 cases were excluded from the study because there was no information from subsequent analysis. They reported a list of 23 individual case reports of VGM. As expected the mortality of these infants was very high.

There were four instances where the infants survived in the postnatal period.

Vintzileos., *et al.* [5] reported a case of VGM where the ultrasonic examination indicated hepatomegaly, cardiomegaly, umbilical vein dilatation and polyhydramnios infant presented with cardiac failure, was treated while it was alive at 10 months of age.

Rizzo [6] published a case report where the infant was born and was waiting for embolization.

Hata [7] in 1988 reported a patient who had successful treatment and survived.

Ishimatsu [8] presented an ante-natal foetus with cardiomegaly which was picked up during routine ultrasound examination and was later diagnosed to have VGM. The cardiac failure was successfully managed, and the malformation was embolized with good outcome.

Evans and Twining [9] diagnosed a patient where the ante-natal ultrasound had diagnosed this cardiomegaly.

Except for these instances all the other cases published in the literature the ultrasound examination diagnosed mainly cardiomegaly and ventriculomegaly. in the postnatal period they either died or were stillborn.

Dr Uggowitzer [10] have published a report on the diagnostic value of echo induced trans cranial doppler sonography and compared the benefits with that of the angiography. These studies were performed in the adults. They concluded that this method was a sensitive investigation to detect AVM's and they go further to reassure that this is safe and effective indicator of Leveovist is effective and safe echo enhancing substance.

The common ultrasonic findings were intra-uterine growth retardation; Mao and Adams [11], ventriculomegaly Reiter [12], Mizejewski [13], Komstoc [14], Lee [15]. In these case reports the main findings during ultrasound examination was ventriculomegaly which led to the further investigation and diagnosis.

In all these reported instances there was mortality mainly because of cardiomegaly and other complications.

Cardiac Failure

Cardiomegaly was a fairly frequently picked up feature Ishimatsu [16] and Paladini [17] Strauss [18], Evans [19], Koven [20], Balister [21]. The cardiomegaly is the most common identifiable feature in the pre-natal ultrasound examination. Because of the large shunting of the arterial blood into the venous system, there is excessive stress on the developing cardiac system. The arterial venous shunt is in effect a massive arterial bleed into the venous system. Therefore, the cardiomegaly is due to the failure of the circulatory system in coping with this massive vascular complication.

The relationship between the malformation and the dynamics of circulation are described in detail by Casikar and Ramaswamy [22,23]. The infants that survived into adult life did not have cardiac failure. This is mainly because the size of the shunt was small, and the infants were able to survive a full term. Vincenzo [24], Leyla [25].

Ultrasound also picks up ventriculomegaly. This occurs mainly because of obstruction to the aqueduct by the dilated vein of Gallen. This is one of the other features which usually draws attention to the possibility of a vascular malformation. VGM is not the primary problem. During the early stages of development the VG is not obstructed by the developing cortex. Arterio- venous shunt presents with large VG dilatation. The arterial component is not often recognised. The various arterial components in this type of Malformations are explained in detail by Casikar [23].

Ventriculomegaly

Ventriculomegaly was reported by Mendelsohn [26], Journal of Ultrasound Medicine [27], Mizejewski [28], Lee [15]. When there is no significant haemodynamic disturbance the survival of the foetus is much better and very often has a good outcome. Au-Yeung [29]. published a case report where the diagnosis was suspected as an anechoic lesion found on pre-natal ultrasound scan. The infant was asymptomatic with normal post-natal growth. There was no evidence of haemodynamic disturbance. This was the only reported incident where the diagnosis of a non-VGM was successfully treated. A good outcome in this instance is mainly because the lesion was not very large. There was no haemodynamic disturbance and the lesion was diagnosed by an ultrasound scan.

Dr Lindegard [30] published a paper on the evaluation of cerebral vascular malformation using trans cranial doppler ultrasound. This study was done in adults in post-natal periods where it was possible to study the velocities of the flow of the fistulae. The pulsatile index could be identified, and a more rational assessment of the haemodynamic damage could be achieved. This method of using trans cranial doppler ultrasound is not possible in the ante-natal period.

The colour doppler ultrasonography has a higher index of recognition of the vascular malformations. However, the reports indicate the diagnosis was mainly related to VG lesion. Smaller lesions of the developing embryo were not identified by this method. Lopez [31], Shanmugam [32], Deloison [33] has published a paper and analysed a high mortality of antenatally diagnosed VGM. The authors concluded that the poor outcomes are influenced by cardiac or cerebral anomalies. They also indicate that isolated VGM tend to have a more favourable outcome. This is not surprising because when there is no cardiac abnormality the flow dynamics of the VG is not very major. This is compatible and therefore has a better outcome. The importance of the flow dynamics in the arterial venous malformation is published by Casikar and Ramaswamy [34,35].

Blaise and associates [36] present their experience in diagnosing and treating 13 children with vein of Galen aneurysmal malformations (VGAM), with an emphasis on possible prognostic indicators, endovascular strategies, factors affecting treatment during the neonatal period, and long-term follow-up. This review identifies those factors that have the most significant prognostic value in determining long-term outcomes in children with VGM. The authors conclude that a coherent policy on such treatment decisions should be developed by the entire group of physicians caring for these children, including neuroradiologists, neurosurgeons, cardiologists, and neonatologists.

MRI

Magnetic resonance imaging has now been adopted in the diagnosis of ante-natal vascular malformation. Glen and Baskovitch [37] have published a review article. In this article they indicate the clinical application of foetal MR imaging. Their studies indicate that the abnormalities of the corpus callosum is associated with vascular complications. They also identify posterior fossa anomalies such as Walker-Warburg syndrome, Chiari and Dandy Walker malformations.

While the MR resonance imaging of the foetal brain is useful in detecting various developmental anomalies, their use in detecting vascular malformation and identifying degree of anomalies of circulation is very limited.

Kosla [38] has discussed the advantages of MR imaging in the diagnosis of VGM. The description is based on their experience of two cases. The authors indicate that ultrasound examination is still the choice of investigation because of its non-invasive character, safety and low cost. The authors indicate that when there is a suspicion of a lesion in the ultrasound examination, magnetic imaging can be a useful complementary method. It is valuable when there is an ambiguous ultrasound picture or there is evidence of hydramnios. When these conditions exist ultrasound, examination is not very useful. The authors also indicate that the MRI examination is not useful as a routine examination because of technical problems and cost involved.

MR examination has certain drawbacks. There is no control over the foetal position. Also, there are multiple artefacts caused by the movement of the foetus. The authors do not recommend breath holding examinations or diaphragmatic compression as it increases the foetal movements. They recommend that MR examination should be conducted in a 1.5 Tessler field with a short spin echo and half ferial angle shot, turbo spin echo and also sequences below to T2 weighted studies. This produces a high resolution of images. The authors conclude that while MR technique is highly valuable in the diagnostics of VGM. This examination gives accurate determination of the ventricular size, elevated ventricular pressure and topographic relationship between pathological vessels and the brain structures. They also indicate that MR examinations give information regarding the presence of infarctions and ischemic areas. This information is not available in ultrasound examination.

The advantage of MR examination however is limited by the availability of the equipment, personnel and the cost of this examination. In the author's view an ultrasound examination is still useful for routine evaluation of ante-natal subjects and MRI examination should be reserved for those instances where the ultrasound examination shows lesions which are not easily diagnosable.

Wagner's [39] have combined the data from Ultrasound and MRI. This method confirms the prenatal ultrasound findings and may identify important secondary complications of the VGAM. The authors are of the opinion that this information will help to monitor progressive heart failure with development of fetal hydrops and hemispheric white matter injuries that are associated with a poor outcome in children with a VGAM.

Doppler Evaluation: Limitations

There are limitations for the use of doppler technology. The most common problem is aliasing. This is a form of an artefact. If the doppler shift frequency exceeds one half of the pulse repetition frequency aliasing occurs. This results in improper doppler shift information. Higher value repetition frequencies permit higher doppler shifts to be detected but this also increases the chances of range ambiguity artefact. Continuous wave doppler instruments do not have this limitation. Unfortunately, they do not have the depth selectivity.

The range ambiguity occurs when the pulse is emitted before all the codes from the previous pulse have been received. When this occurs earlier codes from the previous pulse are simultaneously received with late echoes from the previous pulse. This causes difficulty with deranging process. Unfortunately, the instrument is unable to determine whether the echo is in the early phase or in the deep phase of the examination.

Temporal ambiguity occurs when the doppler colour flow map fails to depict haemodynamic effects with temporal accuracy. Such situations usually occur when the frame rate for colour flow is too slow relative to the circulatory dynamics.

Angle dependency of the doppler is a critical factor in blood flow analysis. When the doppler scanner is used to interrogate the circulatory system, it misses the direction of the flow across the scan lines. The angle of isolation between the flow access and ultrasound beep changes. Concurrently the doppler shifted frequencies progressively decline and may become undetectable.

Mirror imaging is an artefact that occurs in the doppler system. The image of the vessel of the source of doppler shift echoes can be duplicated on the opposite side of the strong reflector. The mirror image can also occur on the opposite side of the baseline. This problem occurs because the beams are focussed and not cylindrical in shape.

There are other less frequent artefacts which can determine the doppler studies. These are speckles. These form of interference affects the scattered sound from the distribution of the scatters created by the etoposides. There may be a constructive interference or a destructive interference. These are various forms of acoustic speckles. Therefore, while the colour doppler measurements have great advantage it is necessary to understand its limitations. Failure to understand this will create major controversies in the results which are published.

The details of the doppler artefacts are published in the new technologies in reproductive medicine and neonatology and gynaecology edited by Cosway., *et al.* Roche – Labarbe [40,41]. They result show that EEG source localization is feasible in neonates. With further development, the technique may prove useful for neurological evaluation of neonates.

Conclusion

In conclusion doppler ultrasonography is still the basis of routine ante-natal examinations. Identification of VGM is usually prompted by cardiac anomalies. A small vascular lesion can be picked up by ultrasound when there is evidence of ventriculomegaly or echogenic defects in the ultrasound examination. Colour doppler studies are superior to routine ultrasound examinations however there are certain drawbacks which need to be identified by the examiner.

MRI imaging is superior to colour doppler studies: however, the use of this is limited by the cost and availability of technical support as a routine examination tool.

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Conflict of Interest

None.

Bibliography

1. Lasjaunias P., *et al*. "The management of vein of Galen aneurysmal malformations". *Neurosurgery* 59.5 (2006): 184-194.
2. Lasjaunias P., *et al*. "Deep venous drainage in great cerebral vein (vein of Galen) absence and malformations". *Neuroradiology* 33.3 (1991): 234-238.
3. Lasjaunias P. "Vein of Galen aneurysmal malformation". Vascular Disease in Neonates, Infants and Children, Springer-Verlas; Berlin (1997): 20-21.
4. Sepulveda W., *et al*. "Prenatal diagnosis of cerebral arteriovenous malformation using color Doppler ultrasonography: case report and review of the literature". *Ultrasound in Obstetrics and Gynecology* 6.4 (1995): 282-286.
5. Vintzileos AM., *et al*. "Prenatal ultrasonic diagnosis of arteriovenous malformation of the vein of Galen". *American Journal of Perinatology* 3 (1986): 209-211.
6. Rizzo G1., *et al*. "Abnormal fetal cerebral blood flow velocity waveforms as a sign of an aneurysm of the vein of Galen". *Fetal Therapy* 2.2 (1987): 75-79.
7. Hata T., *et al*. "Antenatal Doppler color flow mapping of arteriovenous malformation of the vein of Galen". *Journal of Cardiovascular Ultrasound* 7 (1988): 301-303.
8. Ishimatsu J., *et al*. "Evaluation of an aneurysm of the vein of Galen in utero by pulsed and color Doppler ultrasonography. *American Journal of Obstetrics and Gynaecology* 164.3 (1991): 743-744.
9. Evans and Twining. "Case report: in utero diagnosis of a vein of Galen aneurysm using colour flow Doppler". *Clinical Radiology* 44.4 (1991): 281-282.

10. Uggowitzer MM., *et al*. "Value of echo-enhaced Doppler sonography in the evaluation of transjugular intrahepatic portosystemic shunts". *American Journal of Roentgenology* 170.4 (1998): 1040-1046.

11. Mao K and Adams J. "Antenatal diagnosis of intracranial arteriovenous fistula by ultrasoundgraphy". *British Journal of Obstetrics and Gynaecology* 90.9 (1983): 872-873.

12. Reiter A., *et al*. "Prenatal diagnosis of arterio-venous malformation of the vein of Galen". *Journal of Clinical Ultrasound* 14.8 (1986): 623-628.

13. Mizejewski GJ., *et al*. "Combined us of alpha-fetoprotein and ultrasound in the prenatal diagnosis of arteriovenous fistula in the brain". *Obstetrics and Gynaecology* 70 (1987): 452-453.

14. Comstoc CH and Kirk JS. "Arteriovenous malformations, Locations and evolution in the fetalbrain". *Journal of Ultrasound in Medicine* 10.7 (1991): 361-365.

15. Lee W., *et al*. "Atypical presentation of fetal arteriovenous malformation". *Journal of Ultrasound in Medicine* 13.8 (1994): 645-647.

16. Ishimatsu J,. *et al*. "Evalucation of an aneurysm of the vein of Galen in utero by pulsed and color Doppler ultrasoundgraphy". *American Journal of Obstetrics and Gynaecology* 164.3 (1991): 743-744.

17. Paladini D. "Prenatal ultrasound diagnosis of cerebral arteriovenous fistula". *Obstetrics and Gynaecology* 4 (1996): 678-681.

18. Strauss S., *et al*. "Prenatal diagnosis of vein of Galen arteriovenous malformation by duplex sonography". *Journal of Perinatal Medicine* 19 (1991): 227-230.

19. Evans AJ and Twining P. "Case report: in uterodiagnosis of a vein of Galen aneurysm using colour flow Doppler". *Clinical Radiology* 44.4 (1991) 281-282.

20. Koven MB., *et al*. "Foetal intracranial AVM present as enlarged cardiac chamber". *Journal of Ultrasound in Medicine* 12 (1992): 175-183.

21. Ballester MJ., *et al*. "Early prenatal diagnosis of an ominous aneurysm of the vein of Galen by color Doppler ultrasound". *Acta Obstetricia et Gynecologica Scandinavica* 73.7 (1994): 592-5.

22. Casikar V and Ramaswamy GS. "Arteriovenous Malformations of the Brain and Spinal Cord". Published by Wordsworth.

23. Casikar V. Jr of Embryology and Developmental Biology (2017).

24. Vincenzo Suma, Alberto Mrini, Onofrio S Saia, Luca Rigbello. Inner Vision Women's Ultrasound, Nashville, Tennessee, USA (2010).

25. Leyla Karadeniz., *et al*. "Vein of Galen aneurysmal malformation: Prenatal diagnosis and early endovascular management". *Journal of the Chinese Medical Association* 74.3 (2011): 134-137.

26. Mendelsohn DB., *et al*. "In utero diagnosis of vein of Galen aneurysm by ultrasound". *Neuroradiology* 26.5 (1984): 417-418.

27. Editor, Journal of Ultrasound in Medicine. © American Institute of Ultrasound in Medicine. Cover image for Editor-in-Chief: Levon N. Nazarian, MD. Online.

28. Mizejewski GJ., *et al*. "Combined us of alpha-fetoprotein and ultrasound in the prenatal diagnosis of arteriovenous fistula in the brain". *Obstetrics and Gynecology* 70.3 (1987): 452-453.

29. KM Auyeung., *et al*. "Prenatal Diagnosis of Unusual foetalPial Arteriovenous Malformation, A case Report". *Interventional Neuroradiology* 9.2 (2003): 163-168.

30. Lindegaard KF. Evaluation of cerebral AVM's using transcranial Doppler ultrasound". *Journal of Neurosurgery* 65.3 (1986): 335-344.

31. Lopez-Cepero., *et al*. "Prenatal diagnosis of a vein of Galen aneurysmal malformation using colour Doppler ultrasound:a case report". *Boletin De La Asociacion Medica De Puerto Rico* 105.1 (2013): 32-35.

32. Shanmugam S and Bhagavati A. Neuroradiology. 16.4 (2006): 733-756.

33. Deloison GE. *et al*. "Hidden mortality of prenatally diagnosed vein of Galen aneurysmal malformation: retrospective study and review of the literature". *Ultrasound in obstetrics and Gynecology* 40.6 (2012): 652-658.

34. Casikar V., *et al*. "Observations on the flow characteristics of blood flow in Arterio-venous fistula". *Journal of cardio vascular engineering* 10.2 (2010): 66-77.

35. Casikar V., *et al*. "Anatomy and Fluid Dynamics of Cerebral Arteriovenous Malformations". *Annals of Vascular Medicine and Research* 4.2 (2010): 1052.

36. Blaise V Jones., *et al*. "Vein of Galen Aneurysmal Malformation: Diagnosis and Treatment of 13 Children with Extended Clinical Follow-up". *American Journal of Neuroradiology* 23.10 (2002) 1717-1724.

37. Glen OA and Baskovitch J. "Magnetic Resonavce Imaging of the Fetal Brain and the spine". *American Journal of Neuroradiology* 27.8 (2006): 1807-1814.

38. Kosla. Polish Journal of Radiology 78.4 (2013): 88-92.

39. Wagner MW., *et al*. "Vein of Galen Aneurysmal Malformation: Prognostic Markers Depicted on Fetal MRI". *Neuroradiology Journal* 28.1 (2015): 72-75.

40. Cosway. "Reproductive Medicine, Neonatology and Gynaecology, Proceedings of the First International symposium, March Folgaria, Italy (1998).

41. Roche-Labarbe N., *et al*. "High-resolution electroencephalography and source localization in neonates". *Human Brain Mapping* 29.2 (2007): 167-176.

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