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THE HIDDEN MORTALITY OF VEIN OF GALEN ANEURYSMAL MALFORMATION: RETROSPECTIVE STUDY AND REVIEW OF THE LITERATURE

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Abstract:

Objective: To evaluate the prognosis of prenatally diagnosed vein of Galen aneurysmal malformation (VGAM) in a large cohort. To review the literature on prenatally diagnosed VGAM.

Study design: This was a retrospective study of all cases of prenatally diagnosed VGAM managed in our referral center during a 10 years period. VGAM was categorized into isolated forms and forms associated with any other abnormality, based on fetal ultrasound and MRI findings. Poor outcomes comprised termination of pregnancy with confirmation of antenatal findings, perinatal death, and severe cardiac and/or neurological impairment in survivors. The literature was also reviewed and compared for similar cases.

Results: 21 cases of prenatally diagnosed VGAM were managed in our center. Four cases were isolated (19%) and 17 (81%) were associated with other anomalies. There were 9 terminations (43.0%) and 6 neonatal deaths (28.5%). Six children (28.5%) were still alive at last follow-up, 3 of them had an abnormal neurological development. VGAM associated with other anomalies was strongly associated with a poor outcome as compared to isolated forms ($p < 10^{-4}$). One hundred and nine cases from the literature were also reviewed.

Conclusion: Fetuses with prenatally diagnosed VGAM are associated with poor outcomes in the presence of cardiac or neurological anomalies while those with strictly isolated VGAM tend to have a normal outcome, our literature review confirm this finding.

Key words: vein of Galen aneurysmal malformation / prenatal diagnosis/ fetal ultrasound/ fetal MRI/ arterial embolization

Introduction

Vein of Galen aneurysmal malformation (VGAM) is a rare congenital anomaly of the cerebral vasculature, representing less than 1% of all intracranial arteriovenous malformations (1, 2). Nevertheless, VGAM is the most frequent cerebral arteriovenous malformation detected prenatally, representing 30% of all pediatric vascular malformations. VGAM results from an arteriovenous connection between the primitive choroidal vessels and the median prosencephalic vein of Markowski, occurring between the 6th and 11th week of gestation. This leads to abnormal flow, preventing involution of the embryonic vein and subsequent development of the vein of Galen (1, 3-6).

Prenatal diagnosis is usually made during the third trimester, with color Doppler ultrasonography demonstrating turbulent arterial and venous flow within a hypoechogenic structure located in the midline of the posterior part of the 3rd ventricle (3, 7-11). MRI is important to confirm VGAM, to detect associated brain abnormalities, and to rule out differential diagnoses including arachnoid, porencephalic or choroids plexus cysts, pineal tumors, choroid papilloma and intracerebral hematoma (8, 12-14). The cerebral shunt created by the aneurysm can increase cardiac preload and lead to congestive heart failure. Intrauterine ultrasound signs of heart failure, such as cardiomegaly, tricuspid insufficiency, polyhydramnios, pericardial and pleural effusion, edema and ascites carry a poor prognosis, indicating an intractable high-flow anomaly (1, 3, 7, 9, 11, 15, 16).

The value of MRI and ultrasound examination for predicting the outcome of antenatally diagnosed VGAM has rarely been reported (15-17). Here we report our experience in the management of antenatally diagnosed VGAM, and the prognostic value of ultrasound and magnetic resonance imaging.

Patients and methods

We reviewed all consecutive cases of prenatally diagnosed vein of Galen aneurysmal malformation managed in one tertiary referral center between 1999 and 2010. Gestational age assessment was based on crown-rump length (CRL) or certain LMP for all pregnancies. VGAM was suspected during routine antenatal ultrasound examination, based on visualization of an anechogenic or hypoechogenic round or ovoid structure with regular borders located in the midline of the posterior part of the 3rd ventricle. Color Doppler demonstrated the vascular nature of the lesion showing turbulent flow within the malformation (figures 1 and 2).

Confirmatory ultrasound examination was always performed in our center, along with fetal echocardiography and fetal cerebral MRI (figure 3). (Signa; GE, Milwaukee, Wis) in the axial, sagittal and coronal planes,

As VGAM is not associated with chromosomal abnormalities, fetal karyotyping was not routinely proposed. The decision to continue or terminate the pregnancy was made by the parents, after receiving extensive multidisciplinary information on the status and prognosis of their fetus. In France, termination is permitted when a severe fetal abnormality is detected, regardless of gestational age. Full macroscopic and histological examinations of all terminated fetuses were performed to confirm VGAM, to evaluate the severity of brain and/or cardiac involvement, and to detect associated abnormalities.

All ultrasound and MRI examinations were reviewed for this study, including biometric and anatomical findings based on ultrasound reports and stored images. Delivery records were reviewed and pregnancy outcomes were collected. Information on the outcomes of live-born infants was retrieved from pediatric, radiological and neuroradiological records. Information on terminated fetuses and perinatal deaths were retrieved from medical and pathological records. VGAM in fetuses with no associated malformations and/or signs of heart failure was considered “isolated”. VGAM associated with neurological and/or cardiac abnormalities is referred to as “associated VGAM”, since no other associated abnormalities are found with VGAM.

Outcome was considered good if the child had normal cardiac and neurological status at the latest follow-up. Outcome was considered poor in case of termination of pregnancy (with confirmation of antenatal findings), perinatal death, or cardiac and/or neurological

impairment at latest follow-up of living children, we also provide with the outcomes after excluding the TOP which are not option at third trimester in most countries.

We also performed a literature search of the PubMed database from the National Library of Medicine to identify all relevant studies pertaining to the prognosis of antenatal/prenatal detection of VGAM, from 1994 (the year of the Rodesh study (17) and one year before 1995 Sepulveda's comprehensive review of the literature (2)) onwards. A database was compiled and subsequently reviewed. We reviewed the articles, and excluded those that did not meet our eligibility criteria. Retrospective studies, case reports and expert opinions were included. Common reasons for exclusion were articles in an unknown language or that did not focus sufficiently on the antenatal prognosis of VGAMs.

Stata 9.2 for Windows (StataCorp LP, TX 77845 USA) was used for statistical analyses. All tests were two-tailed and p values of < 0.05 were considered to denote statistical significance.

Results

During the study period, 21 cases of prenatally diagnosed VGAM were managed in our unit. The socio-demographic details of the study population are shown in *Table 1*.

Fetal morphological cerebral and cardiac ultrasound findings and fetal cerebral MRI findings were available in all cases and are summarized in *Table 2*. The most common finding associated with a poor outcome was cardiomegaly and ventriculomegaly (67% and 39% respectively). Noteworthy, in three cases, gyration was considered normal on US whereas MRI found abnormal gyration.

The outcomes of the 21 pregnancies are shown in the flow chart (*Figure 4*). The parents chose to terminate the pregnancy in 9 cases (43.4%) after parents' counseling. On the twelve born children, 3 (25%) had good outcomes (14.3% of the whole series) and 9 (75%) had poor outcome (43.4% of the whole series) with 6 post-natal deaths, and 3 cases of moderate to severe mental retardation in living children.

Women chose to terminate the pregnancy in 9 cases. Pathological examination confirmed the prenatally diagnosed anomalies in all cases (*Table 3*). The remaining 12 pregnancies went to term. There were 6 neonatal deaths (*Table 4*). Autopsy was declined in four of these cases. One of the two autopsied children had no associated abnormalities. He had received embolization at the age of 3 months, but the coil migrated through the venous system, leading to fatal thrombosis of the venous system.

Neonatal disorders and causes of death are summarized in *Table 4*. Only two of the six dead children required embolization: the aforementioned child, and another child who had two emergency embolization procedures at 8 and 15 days of life for persistent cardiac dysfunction. He died at age 7 weeks, of cardiac failure. The other four neonates died of cardiac failure, without undergoing embolization.

Among the 6 children who were still alive at latest follow-up, 3 were considered to have isolated VGAM prenatally. These 3 children received embolization between 5 and 18 months of life while they had no cardiac or developmental abnormalities. The three children are alive and well at the ages of four, three and one year. The other three children were considered to have associated abnormalities prenatally. One child had a prenatal diagnosis of hydrocephalus. He received embolization on day 18 of life and showed severe developmental

and mental retardation at 2 years. The other two children with associated abnormalities prenatally (mild ventriculomegaly and macrocephaly in one case each) had moderate mental retardation at 6 and 18 months of life, respectively.

The prognostic value of prenatal ultrasound findings is summarised in *Table 5*. Associated abnormalities were strongly associated with poor outcome ($p < 10^{-4}$) compare to isolated VGAM.

Table 6 presents the data from 109 cases of antenatally diagnosed and published VGAM from 1994 to 2011. In this section, the prognosis factors of patients with antenatally diagnosed VGAMs were examined and are summarized in this table. After exclusion of TOPs, the total number of cases with known outcome was 91 and the outcomes were: 49 (54%) perinatal deaths, 29 (32%) alive and well and 13 (14%) alive with mental retardation (details in Table 6).

Discussion

The prognosis of prenatally diagnosed VGAM was far poorer in this series than generally reported (15, 17). Outcome was particularly poor when associated anomalies such as cerebral defects or signs of cardiac dysfunction were detected prenatally. These new data are important for all those concerned by the antenatal management of VGAM.

Transarterial endovascular embolization was thought to have improved the prognosis of VGAM. In the study by Rodesh et al (17), 12 of 18 children with prenatally diagnosed VGAM were embolized, and 8 (67%) of these 12 children had normal neurological outcomes. In contrast, only 3 children (14.3%) are alive and well in our series of 21 cases. It is particularly noteworthy that VGAM was the only prenatally detected abnormality in all 3 of these children. This difference in outcomes between the two studies may be explained by the fact that Rodesh's series included no fetuses with associated neurological defects, possibly because they were considered to have too poor a prognosis for referral to a neuroradiological team. Like Rodesh and others (9, 15-18), we found that cardiac anomalies as simple as antenatal cardiomegaly were strongly associated with poor outcome.

This is the largest series of prenatally diagnosed VGAM published to date. Importantly, we included all cases managed in our center, minimizing any selection bias and thus revealing the previously unrecognized morbidity and mortality of prenatally diagnosed VGAM. Indeed, most previous studies, conducted mainly in neuroradiology units, included only patients considered eligible for embolization.

In 1997, Yuval et al (7) proposed a number of prognostic features, including fetal hydrops, anatomical abnormalities, cardiomegaly, dilated vena cava or jugular vein, retrograde aortic flow, and multiple feeding arteries. However, our review is more pessimistic especially for some prognosis factors like multiple feeding vessels, dilated jugular vein, retrograde aortic flow or cardiomegaly which seem to be associated with a poorer prognosis. Our report suggests that, at least for antenatal counseling, Yuval's prognostic factors may be simplified: good outcome may be expected if no cardiac or neurological defects are present prenatally, whereas outcome is likely to be poor if any such defects are detected.

Indeed, associated abnormalities such as cardiac dysfunction turn VGAM into a life-threatening condition requiring emergency embolization in the first days of life. Embolization in neonates, and especially in hemodynamically unstable neonates, is more difficult and carries a high risk of complications and death (18). Intracranial bleeding is a major risk, and

persistent venous insufficiency following embolization may cause hydrocephalus, progressive parenchymal atrophy and calcification (6, 15, 16, 18). Our study thus confirms that prenatal signs of cardiac dysfunction are an ominous finding in fetuses with VGAM.

Few other studies (2, 7, 14, 19) proposed a number of prognostic features, most of the cases of antenatal VGAM are short reports or case reports they are summarized in table 6. Our review of the literature find also a poor outcome of antenatally diagnosed VGAM since about 30% of the patient are alive without mental retardation. The prognosis factors suggested in the literature are much more detailed than we did, but they all reports a poor prognosis of any cardiac associated sign (like cardiomegaly, dilatation of the right atrium and superior vena cava, tricuspid valve regurgitation, dilated jugular vein, retrograde aortic flow, hydrops...) or any cerebral damage. However the angioarchitecture of the malformation seems also to be an ominous prognostic factor in this review, choroidal type consists of multiple direct, high-flow shunts into choroidal tributaries draining into the midline venous aneurysm and when the arteriovenous fistula is in the wall of the dilated vein itself, this is referred to as the mural type. The mural angioarchitecture is lower flow with fewer fistulas and presents later in childhood which seems to be of better prognosis. However the angioarchitecture of the VGAM is hardly evaluated prenatally. Some studies reported that the angioarchitecture could be depicted prenatally with 3D CPA (10) or with Magnetic resonance angiography (13) but the diagnosis is easier post natally.

Regarding the limitations of our study, the small size of the fetal population is due to the rarity of VGAM and the fact that it is usually diagnosed postnatally. In addition, all the patients had been referred to our tertiary center, raising the possibility that they represented the most severe cases. However, this is unlikely, as VGAM is generally considered to require delivery in a tertiary care facility with a specialized neonatal care unit. Finally, although our simple classification of antenatal VGAM as “isolated” or “associated” yielded interesting information, it might be challenged in future studies; even if no signs of severe intrauterine cardiac failure are found, fetuses with VGAM can develop significant cardiac impairment shortly after birth (9, 18). Another limitation of our study is that it provides an in-depth literature review of VGAM however it is not a systematic review, because articles were not scored on a formal grading scale of inclusion and exclusion criteria. In this review most of cases are cases reports with all the bias and limitations of this type of paper. However, given

the rarity of VGAM, this paper provides valuable information regarding the natural history, treatment, and outcomes of this condition.

Conclusion

Our results suggest that prenatally diagnosed aneurysm of the vein of Galen carries a grim prognosis, except when no signs of associated cardiac failure or neurological abnormalities are present. Close follow-up and a thorough radiological search (ultrasound and MRI) for associated anomalies form the basis of parental counselling. The results of the study may be helpful for prenatal counselling; specifically preparing parents for the outcomes including short- and medium-term morbidity associated with VGAM. The validity of our simple and pragmatic prognostic classification fits the literature data though it should be prospectively validated.

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Figure 1: Anechoic, round, median lesion (star) of the brain on ultrasound examination

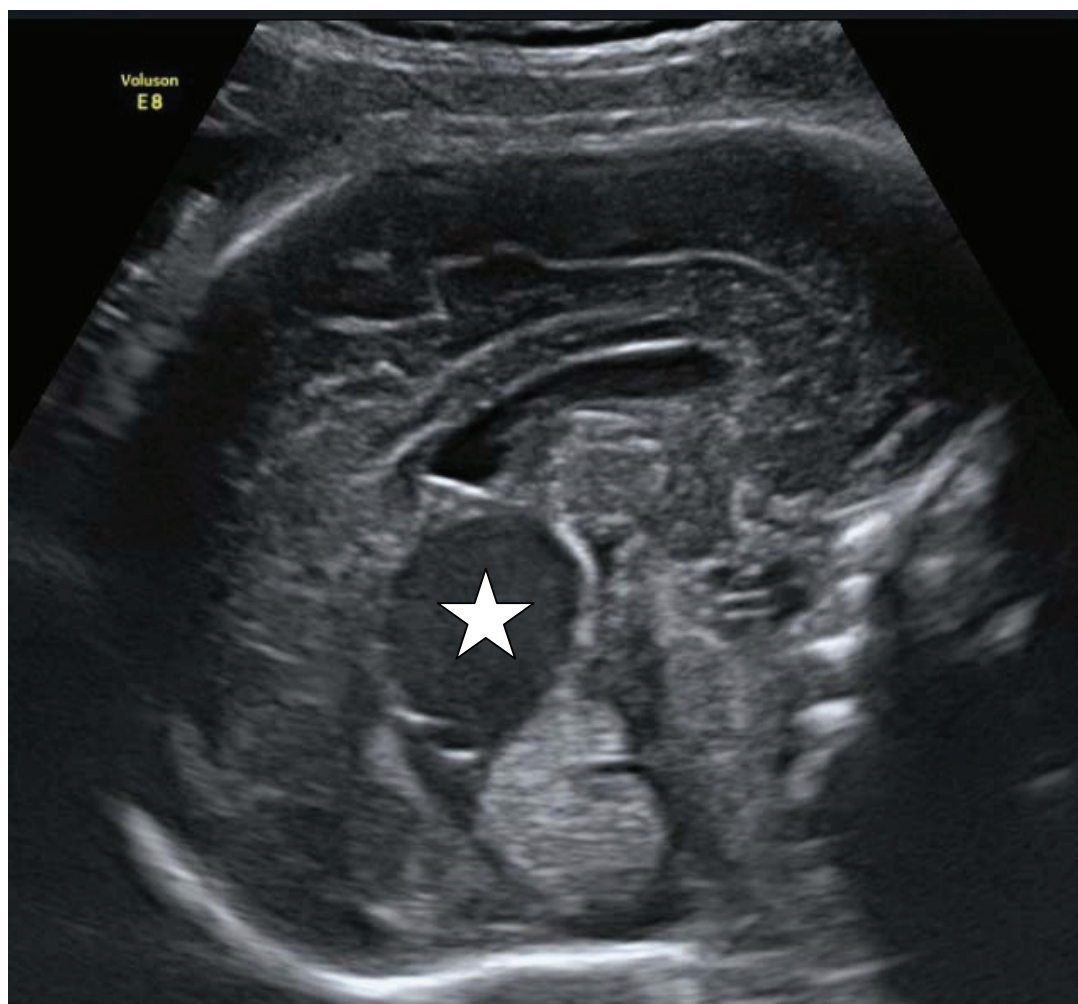


Figure 2: Color Doppler Imaging demonstrating the vascular nature of the lesion and showing turbulent flow.

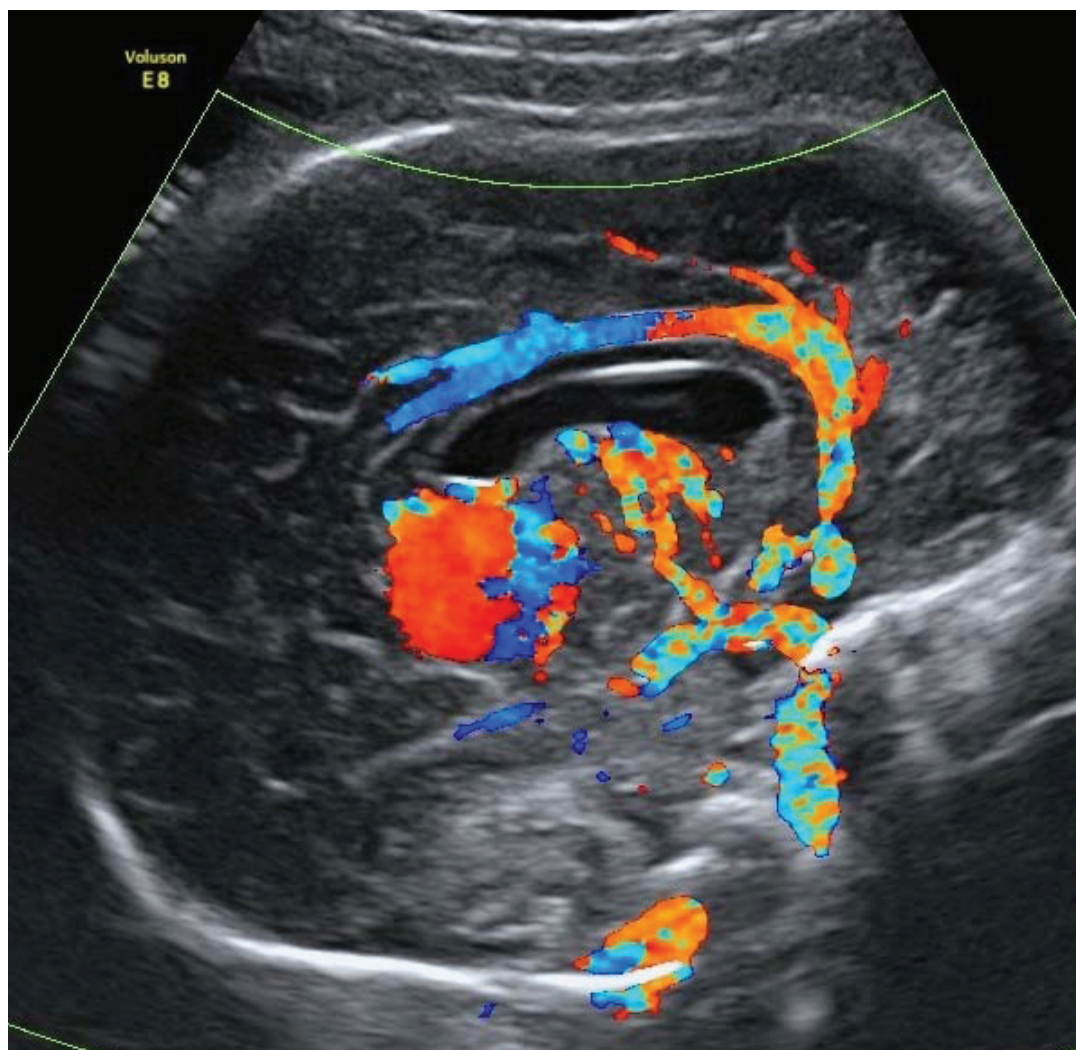


Figure 3: T2-weighted Fetal MRI images demonstrating VGAM (star) on sagittal (a) and axial (b) planes.

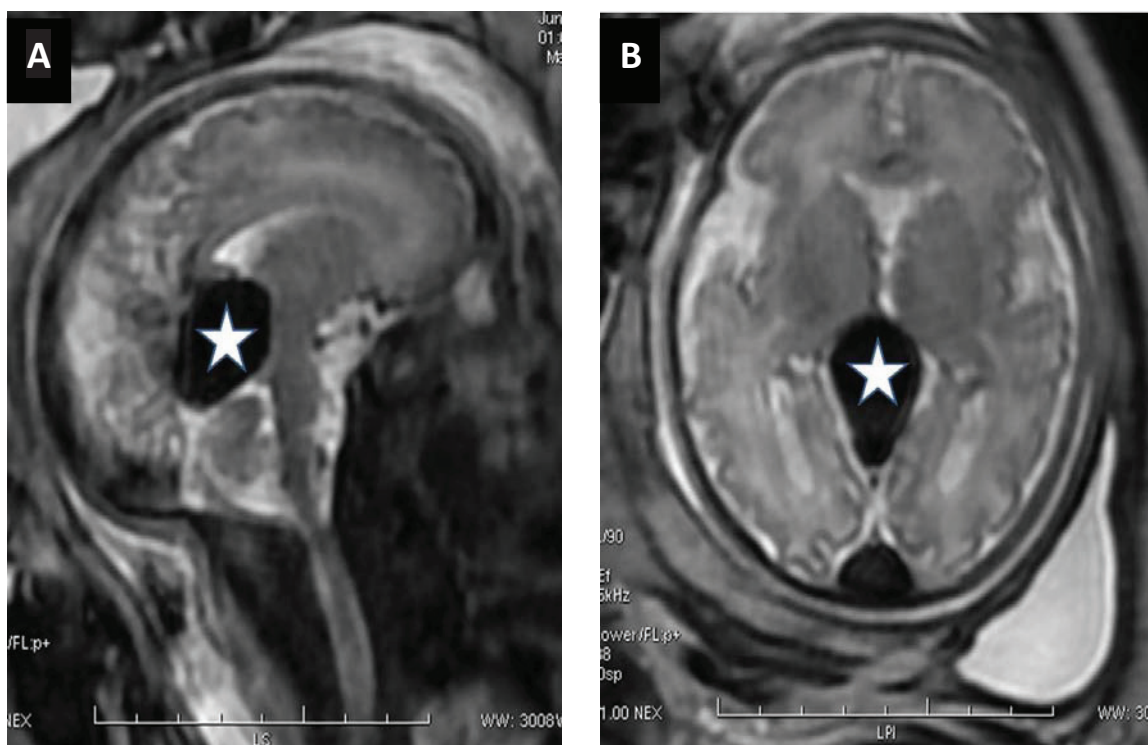
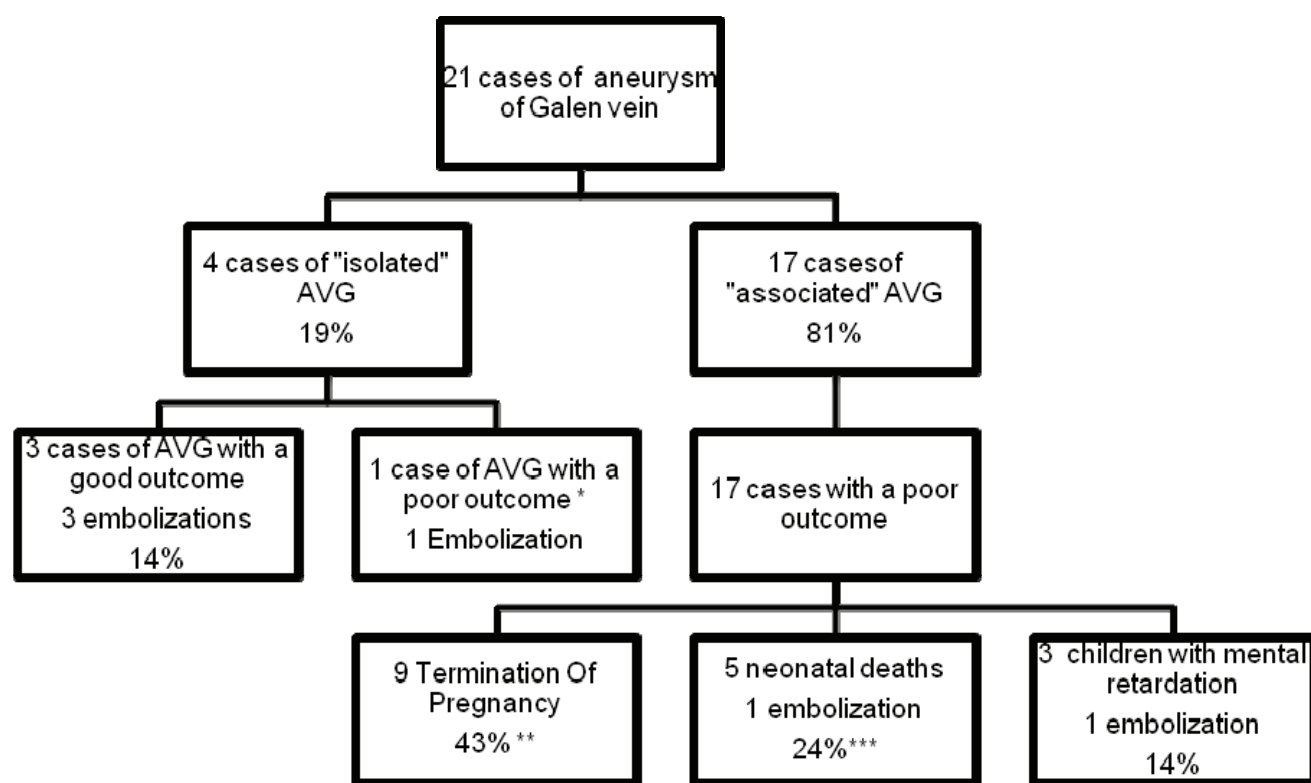


Figure 4: Outcomes of 21 cases of vein of Galen aneurysmal malformation (GVA)



*: This child had embolization at age 3 months, but the coil migrated through the venous system, causing fatal thrombosis of the central venous system of the brain

**: Details in table 3.

***: Details in table 4.

Table 1: Characteristics of the study population

	Overall (n=21)	TOP (n=9)	Neonatal death (n=6)	Alive at last follow-up (n=6)
Maternal age (years) +/- SD	31.5 +/- 5.8	29 +/- 4.1	34.5 +/- 5.6	32.2 +/-7.3
Median [range] gravidity/parity	3/1	1/0	3/2	3/2
Gestational age at diagnosis (weeks) +/- SD	32.8 +/- 2.1	33.4 +/-1.7	32.8 +/-1	32.2 +/-3.9
Gestational age at MRI (weeks) +/- SD	34.6 +/-2.4	34.7 +/-2.1	34.8 +/-0.8	34.1 +/-3.4
Gestational age at birth (weeks) +/- SD	38.7 +/-2.2	35.7 +/-2.1	38 +/- 1.4	39.3 +/-0.5
Birth weight (g) +/- SD	3096 +/-445	2700 +/-90	3120 +/-260	3530+/-450
Birth head circumference (cm) +/-SD	33.9 +/-2.8	31.5 +/-1.6	35.2 +/- 1.7	35.6 +/-3.1

SD = standard deviation

Table 2: Associated antenatal ultrasound and MRI findings in fetuses with aneurysm of Galen vein, according to outcome

<i>Findings</i>		<i>Poor outcome after exclusion of TOP (n=9)</i>	<i>Poor outcome (n=18)</i>
Cerebral ultrasound and MRI abnormalities	<i>Ventriculomegaly</i>	2	7 (39%)
	<i>Hydrocephaly</i>	0	2 (11%)
	<i>Polymicrogyria</i>	0	2 (11%)
	<i>Cortical thickening</i>	0	2 (11%)
	<i>Porencephaly</i>	0	1 (5%)
	<i>Schizencephaly</i>	0	1 (5%)
	<i>Periventricular leukomalacia</i>	0	1 (5%)
	<i>Intracerebral hemorrhage</i>	0	1 (5%)
Cardiac abnormalities	<i>Cardiomegaly</i>	5	12 (67%)
	<i>Tricuspid regurgitation</i>	2	5 (28%)
	<i>Pericardial effusion</i>	1	2 (11%)
	<i>Vena cava dilatation</i>	0	2 (11%)
	<i>Hydrops</i>	0	1 (5%)

Table 3: Details of terminated pregnancies and autopsy findings

Case number	Antenatal classification	Ultrasound and MRI abnormalities	Cardiac abnormalities	Autopsy findings
1	Associated VGAM	Hydrocephaly	Cardiomegaly	Triventricular cerebral dilatation with ischemic brain lesion and cardiomegaly
2	Associated VGAM	Porencephaly and ventriculomegaly	Major cardiac dysfunction	Hydrocephaly, cerebral necrosis lesions and cardiac lesions
3	Associated VGAM	Ventriculomegaly	Cardiac dysfunction with cardiomegaly and subcutaneous edema	Intracerebral hemorrhage and cardiomegaly.
4	Associated VGAM	Micropolygyria and ventriculomegaly	Cardiomegaly and tricuspid dysfunction	Extensive cerebral necrosis and cardiomegaly
5	Associated VGAM	Periventricular leukomalacia	Fetal hydrops with cardiomegaly	Fetal hydrops and extensive ischemic brain lesion
6	Associated VGAM	Schizencephaly, ventriculomegaly and IUGR	Cardiomegaly and tricuspid insufficiency	Extensive cerebral necrosis, cardiomegaly and IUGR
7	Associated VGAM	Hydrocephaly	Cardiomegaly	Micropolygyria, hydrocephaly, cardiomegaly
8	Associated VGAM	Ventriculomegaly	Cardiomegaly	Intracerebral hemorrhage, fetal hydrops
9	Associated VGAM	Porencephaly and ventriculomegaly	Cardiomegaly	Intracerebral necrosis and hemorrhage, cardiomegaly

IUGR = intrauterine growth restriction

Table 4: Details of the neonatal abnormalities of the 6 children who died, and causes of death (VD: vaginal delivery; C: Cesarean section)

	<i>Antenatal classification</i>	<i>Delivery</i>	<i>Neonatal defects</i>	<i>Time of death</i>	<i>Cause of death</i>
1	Associated VGAM	VD	Respiratory distress and cardiac failure	41 days	Cardiac failure
2	Associated VGAM	C	Major cardiac failure	6 days	Cardiac failure
3	Associated VGAM	C	Cardiac failure	3 days	Cardiac failure
4	Associated VGAM	VD	Cardiac failure	2 days	Cardiac failure
5	Isolated VGAM	VD	No symptoms	98 days	Coil migration with cerebral thrombosis after embolisation
6	Associated VGAM	VD	Major cardiac failure	10 hours	Cardiac failure

Table 5: Relation between ultrasound and MRI parameters and outcome

<i>Antenatal findings</i>		<i>Total</i>	<i>Poor outcome</i>	<i>Good outcome</i>	<i>p</i>
<i>Any antenatal anomaly</i>	Present	17	17	0	$p < 10^{-4}$
	Absent	4	1	3	
<i>Cerebral anomaly</i>	Present	11	11	0	$p < 0.05$
	Absent	10	7	3	
<i>Cardiac anomaly</i>	Present	12	12	0	$p < 0.05$
	Absent	9	6	3	

Table 6: Summary of studies examined the correlation between prenatal ultrasound markers and postnatal outcome.

Reference	No. of antenatal cases	Study Design	Prenatal Ultrasound findings predictive of poor outcome	Prenatal Ultrasound findings predictive of good outcome	Outcome (n)	Comments
Rodesh 1994(17)	18	RS	- destruction of the brain parenchyma -Sign of CHF		6 alive without MR 4 still under treatment (2 with moderate MR) 2 alive with moderate MR 4 deaths from CHF 1 TOP 1 LFU	pediatrics' serie
Sepulveda 1995(2)	1 CR and review of 24 CR	CR	-Cardiomegaly -Jugular dilatation -Ventriculomegaly -Polyhydramnios - hepatomegaly -Ascite -IUGR		14 deaths 5 alive 2 TOP 2 IUFD 2 without outcome	1 CR and review of 24 CR
Campi 1996 (14)	1	CR	-Choroidal angioarchitecture -Brain damages	-Mural angioarchitecture	Alive without MR	
Goelz 1996(20)	2	CR	-Hydrocephalus -Sign of CHF -Huge shunting of blood flow in the prosencephalic vein		2 neonatal death	
Mai 1996(21)	1	CR		-No sign of CHF -no associated anomaly	Alive at 6 months after embolization	
Yuval	2	CR	-Anatomical brain	- Stenotic or normal-to-minimally dilated	1 alive	

1997(7)			changes	straight sinus	1 neonatal death from CHF	
			-Hydrops	-No intracranial venous anomaly except VGAM		
			- Retrograde aortic flow			
			-Multiple (5 or more) feeding vessels	-Preserved brain anatomy		
			-Dilated jugular vein and/or IVC	-Two or less feeding arteries		
			- Dilated drainage tract	-No evidence of high output state		
			-Cardiomegaly			
Pilu	4	CR	-Hydrops	-Normal structure brain	2 neonatal deaths	
1997(22)			-Significant ventriculomegaly	-Small lesion	1 TOP	
			-cerebral edema		1 alive without MR	
Paumier	2	CR	-Hydrops	-no CHF sign	1 TOP for hydrops	
1998(23)			-Prenatal signs of CHF		1 IUFD from CHF	
Campi	1	CR	No	No	1 death after surgery	3 CR (only one antenatal diagnosis)
1998(24)						
Heling	1	CR	No	No	1 death at 5 months from CHF	Use of 3D power Doppler
2000(25)						
Bahlmann	1	CR	No	No	Alive without MR	Use of 3D power Doppler
2000(26)						
Kurihara	1	CR	-Brain injury		1 death from CHF	
2001(13)			-Severity of CHF			
			-Volume of arterio-venous shunting: feeding arteries and venous drainage			
Brunelle		EO	-Fetal hydrops	-No cardiac failure	No	
2003(19)			-Sign of CHF	-No brain atrophy		
			-Vascular steal with high resistive index in the sylvian arteries	-Low resistivity index of the sylvian artery		

			-Brain atrophy -Complex angioarchitecture			
Hartung 2003 (27)	1	CR	-Sign of CHF		1 TOP	Early diagnosis at 22 weeks
Sepulveda 2003 (3)	1	CR	-Neck enlarged vessels -Cardiomegaly -Multiple feeding vessels	-No other cerebral structural anomalies	Alive at 3 months	Use of 3D power Doppler angiographic imaging modality
Has 2003(28)	1	CR	-Hydrocephalus, -Cardiomegaly -Dilated jugular veins	No	Death after 3 days from CHF	
Ruano 2003(10)	3	CR	-Fetal heart failure -Porencephaly		1 child alive TOP 1 death	Use of 3D power Doppler
Gerards 2003(29)	2	CR	-Structural brain damage -Dilated drainage tract -Multiple (≥ 5) feeding vessels -Hydrops -Dilated jugular vein -Dilated inferior vena cava -Retrograde aortic flow -Cardiomegaly		1 child with moderate MR 1 neonatal death from CHF	Almost the same prognosis factors as Yuval
Messori 2003(12)	1	CR	No	no	1 alive child without MR	
Paternoster 2003(11)	1	CR	-Sign of CHF		1 IUFD	
Hirose	1	CR	-Sign of CHF (cardiomegaly,		1 neonatal death from	Huge aneurysm

2003(30)			dilatation of the right atrium and superior vena cava, tricuspid valve regurgitation) -Hydrocephalus -Huge aneurysm		CHF	
Fullerton	5	RS	-CHF		No details	
2003(31)			-Choroidal angioarchitecture			
Beucher	1	CR		-No sign of CHF	1 death from CHF	
2005(1)				-No associated cerebral anomaly		
				-Normal Gyration		
Gailloud		EO	-brain atrophy			
2005(32)			-CHF			
Doyle	1	CR	-Signs of CHF		1 neonatal death from CHF	
2005(33)			-Brain injury -multiple arterial feeding vessels			
Vijayaraghavan	1	CR	-Distortion of the cerebral architecture		1 TOP	Thrombosed VGAM
2006(34)						
Cherif	1	CR	No	No	1 death from CHF	Late onset of CHF without antenatal sign
2007(35)						
Santo	2	CR	- Choroidal angioarchitecture	-Mural angioarchitecture	2 neonatal deaths	
2008(9)			- Associated cerebral injury -Intrauterine signs of cardiac failure			
Geibprasert	8	RS	-Arterial steal -Mural angioarchitecture with multiple holes -Choroidal angioarchitecture	-Mural angioarchitecture with single hole	No details	Pre and pos- natal diagnosis (9/20 of good outcome in the whole serie)
2009 (18)						

			-Multiorgan failure -Brain parenchymal changes			
Gun 2009 (36)	1	CR	-Volume of the aneurysm		1 severe MR	Evaluation of 3D US for aneurysmal volume calculation
McSweeney 2010(37)	7	RS	No	No	No details	Pre and postnatal series (14/26 good outcome in the whole series)
Heuer 2010(38)	6	RS	-Choroidal angioarchitecture -Parenchymal brain damage -Signs of progressive cardiac dysfunction	-mural angioarchitecture	2 alive children without MR 2 alive children with MR 2 death	Pre and postnatal diagnosis
Kalra 2010(39)	1	CR	No	No	1 death from CHF	
Berenstein 2011(40)	5	RS	-Cardiomegaly -Signs of CHF -Choroidal angioarchitecture -Brain parenchymal abnormalities		3 alive children without MR 2 alive children with MR	Only outcomes after embolization are treated in this article
Our serie	21	RS	-Any cerebral associated anomaly -Cardiac anomaly	No associated anomaly	3 alive children without MR 3 alive children with MR 6 deaths 9 TOP	
TOTAL	130	25 CR 6 RS 2 E0			44 deaths 29 alive without reported MR 13 alive with MR	

16 TOP

5 IUFD

TOP : Termination Of Pregnancy

IUFD: Intra Uterine Fetal Death

CHF: Congestive Heart Failure

MR: Mental Retardation

LFU: Lost to Follow-Up

RS: Retrospective Study

CR: Case Report

EO: Expert Opinion