

Prenatal diagnosis of Galen vein aneurysm: when to deliver?

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Abstract

Objective: Prenatal diagnosis of Galen vein aneurysm using color Doppler has become relatively easy. Management and timing of delivery is however challenging because of cardiac function deterioration. Ultrasound follow-up is essential to detect signs of fetal cardiac failure which must be weighed against risks of prematurity. Postnatal deterioration of the vital signs can follow a short period of improvement after birth. This report illustrates the many pitfalls that could affect the final outcome, raising the issue of the proper timing for delivery.

Case: A 23-year-old G1P0 spontaneous pregnancy was referred at 28 WA for a Galen vein aneurysm. The ultrasound finding included a 35 by 14 mm vascular structure with a turbulent flow, occupying the central part of the head. After 48 hours in-hospital surveillance through non-stress tests twice daily, she had a cesarean section due to a persistent non-reactive fetal heart rate, with absent variability. Unfortunately, despite a stable state for the first ten days, the baby's cardiac function deteriorated abruptly on day 11, and the baby died of heart failure despite medical management.

Conclusion: The timing of delivery in cases with Galen vein aneurysm is challenging mainly in the case of premature babies, as the prognosis is globally bad. Postnatal deterioration of the vital signs can follow a short period of improvement after delivery.

Keywords: Fetal, Galen vein aneurysm, malformation, prematurity, ultrasound.

Introduction

Galen vein aneurysm is a rare occurrence involving cerebral vessels and leading to a high output cardiac failure, potentially leading to fetal demise. We report a case that highlights the difficulty in the clinical management of this entity.

Case Report

A 23-year-old G1P0 spontaneous pregnancy was referred at 28 WA for a Galen vein aneurysm. The ultrasound finding included a tubular irregular vascular structure (35×14 mm in diameter) with a turbulent flow, occupying the central part of the head (**Fig. 1**).

Biometry was around the 20th percentile. Cardiac overload was evident, with a spherical shape heart, dilated neck vessels, and tricuspid regurgitation. At 33 weeks, IUGR was more obvious, reaching the 10th percentile for all the biometric parameters. Umbilical artery Doppler then displayed an absent diastolic flow; and ductus venosus revealed an abnormal a wave. Mid-cerebral artery Doppler was measurable despite the aneurysm and remained in the normal range. After 48 hours in-hospital surveillance by non-stress tests twice daily, she had a cesarean section due to a persistent non-reactive fetal heart rate, with a pathologically reduced variability. The baby weighed 1800 g and was doing relatively well, with a 5-minute Apgar score of 9.

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As the baby was stable for the first 48 hours, occlusion of the aneurysm by interventional radiology was postponed until reaching a weight of 2500 g. This decision was based on literature review—expert opinions from centers having a relatively extensive experience in cases of Galen vein aneurysm—and furtherly based on the preference of our radiology team. Unfortunately, despite a stable neonatal condition for the first ten days, the baby's cardiac function deteriorated abruptly on day 11, and the baby died of heart failure despite medical support. This case raises the question regarding the timing of delivery and scheduling the interventional radiology procedure, with both prenatal and postnatal outcomes being at risk for these babies. This case report has been approved and registered by our institution's review board under the number CEHDF-899.

Discussion

Despite its rarity, Galen vein aneurysm represents the most frequent cerebral arteriovenous malformation detected prenatally, and approximately 30% of all pediatric vascular malformations. The prognosis depends on gestational age at diagnosis and aneurysm size.^[1] Tricuspid regurgitation, major brain lesion and volume of the lesion on MRI are also associated with a poor outcome.^[2] Prognosis also depends upon prematurity as most babies are delivered because of the risk of heart failure.

Some cases present regression following delivery, suggesting a more conservative approach after delivery.^[3] In our case, despite a relatively favorable postnatal outcome, a rapid deterioration of the cardiac status occurred after 11 days. A similar case has been reported by Cherif et al., who evidenced that after an initial period of improvement, the cardiac status deteriorated suddenly on day 14, which led to fetal death on day 36 despite medical treatment; the authors concluded that heart failure can occur much after the first days of life.^[4] Embolization is a therapy of choice; the results are variable as a few authors report high failure rate and neonatal death.^[1,5]

Prognostic features on ultrasound have been reported, and include mapping of intracranial feeding arteries, assessment and measurement of flow in the straight sinus, existence of 'steal' retrograde aortic

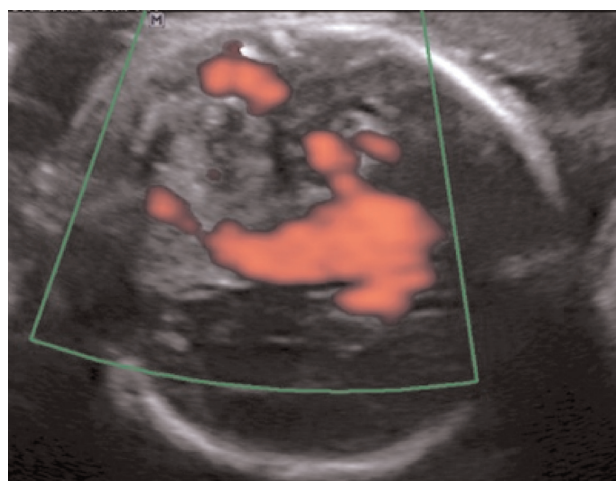


Fig. 1. Cerebral central hypoechoic vascular structure, with turbulent flow.

flow, and the appearance of high-output cardiac state.^[6] The related outcome appears poor, with only about 30–50% of patients diagnosed with the condition being alive without mental impairment.^[1,2] Deciding for delivery to allow radiology intervention appears to have a limited impact due to the existing serious prognosis involved, and the timing of this decision can be challenging.

Conclusion

The timing of delivery in cases with Galen vein aneurysm is challenging mainly in the case of premature babies, as the prognosis is globally bad. Postnatal deterioration of the vital signs can follow a short period of improvement after delivery.

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References

1. Deloison B, Chalouhi G, Sonigo P, Zerah M, Millischer E, Dumez Y, et al. Hidden mortality of prenatally diagnosed vein of Galen aneurysmal malformation: retrospective study and review of the literature. *Ultrasound Obstet Gynecol* 2012;40:652–8. [PubMed] [CrossRef]

2. Paladini D, Deloison B, Rossi A, Chalouhi GE, Gandolfo C, Sonigo P, et al. Vein of Galen aneurysmal malformation (VGAM) in the fetus: retrospective analysis of perinatal prognostic indicators in a two-center series of 49 cases. *Ultrasound Obstet Gynecol* 2017;50:192–9. [[PubMed](#)] [[CrossRef](#)]
3. Cerbo RM, Cabano R, Radicioni M, Stronati M. Cerebral arteriovenous shunts regression in two preterm newborns with heart failure in utero. *Am J Perinatol* 2009;26:637–9. [[PubMed](#)] [[CrossRef](#)]
4. Cherif A, Néji K, Sebaï L, Ben Jemaa W, Bellagua I, Jebnoun S, et al. Vein of Galen aneurysmal malformation: a neonatal case with unusual evolution. [Article in French] *Arch Pediatr* 2007;14:893–6. [[PubMed](#)] [[CrossRef](#)]
5. Santo S, Pinto L, Clode N, Cardoso E, Marques JP, Melo A, et al. Prenatal ultrasonographic diagnosis of vein of Galen aneurysms – report of two cases. *J Matern Fetal Neonatal Med* 2008;21:209–11. [[PubMed](#)] [[CrossRef](#)]
6. Yuval Y, Lerner A, Lipitz S, Rotstein Z, Hegesh J, Achiron R. Prenatal diagnosis of vein of Galen aneurysmal malformation: report of two cases with proposal for prognostic indices. *Prenat Diagn* 1997;17:972–7. [[PubMed](#)] [[CrossRef](#)]

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