



# Vein of Galen Aneurysmal Malformation in Neonates Presenting With Congestive Heart Failure

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## Abstract

The authors report the case of a neonate presenting with signs of a congenital cardiac disease. Echocardiography showed a structural normal heart, right-to-left ductal flow, a dilated superior caval vein, and reversed diastolic flow in the proximal descending aorta. Brain magnetic resonance imaging showed a vein of Galen arteriovenous malformation. This highlights the importance of considering an intracranial cause in the differential diagnosis of neonatal congestive heart failure.

## Keywords

neonate, vein of Galen malformation, cardiac failure

Received August 21, 2015. Received revised October 14, 2015. Accepted for publication November 27, 2015.

Vein of Galen aneurysmal malformation (VGAM) is a rare cause of congestive heart failure in the neonatal period and carries a high mortality rate. It is a difficult diagnosis because the clinical picture often presents with symptoms suggesting that the patient has a congenital cardiac malformation.<sup>1,2</sup> Patients are subjected to high-risk procedures such as cardiac catheterization to establish a diagnosis. These investigations often have negative findings and can delay correct diagnosis.

## Case Study

A term boy was born by cesarean section for fetal distress after an uncomplicated pregnancy. Apgar scores were 6 and 6 at 1 and 5 minutes, respectively. At 30 minutes after birth, he became cyanotic and needed extra oxygen. Physical examination revealed a cardiac murmur. A diagnosis of congenital heart disease was suspected, and treatment with prostaglandin E1 was initiated. The neonate was intubated and ventilated, and he was transferred to our neonatal intensive care unit. On admission, he had tachypnea and cyanosis, and his oxygen saturation was 94% with a fraction of inspired oxygen of 70%. Physical examination showed a loud systolic cardiac murmur and prominent venous neck pulsations, decreased peripheral pulsations, and hepatomegaly. Neurological examination showed an agitated neonate with a head circumference of 37 cm (90th

percentile), a fontanel in level, a cephalohematoma, and a generalized hypertonia. Auscultation of the fontanel was not performed. An aortic coarctation was suspected, prostaglandin E1 was continued, and circulatory support with dobutamine was started.

Chest X-ray showed an enlarged heart. Echocardiography showed a structurally normal heart with a dilated right ventricle with suprasystemic pulmonary arterial pressures based on Doppler velocity measurements of a moderate tricuspid valvular insufficiency. There was a normal aortic arch without apparent coarctation. An open ductus arteriosus was present with systolic right-to-left ductal flow. Within the proximal descending aorta, there was substantial diastolic retrograde flow into the carotid arteries. Finally, a dilated superior caval vein was observed (Figure 1). Based upon these findings, a low resistance cerebral arteriovenous malformation was

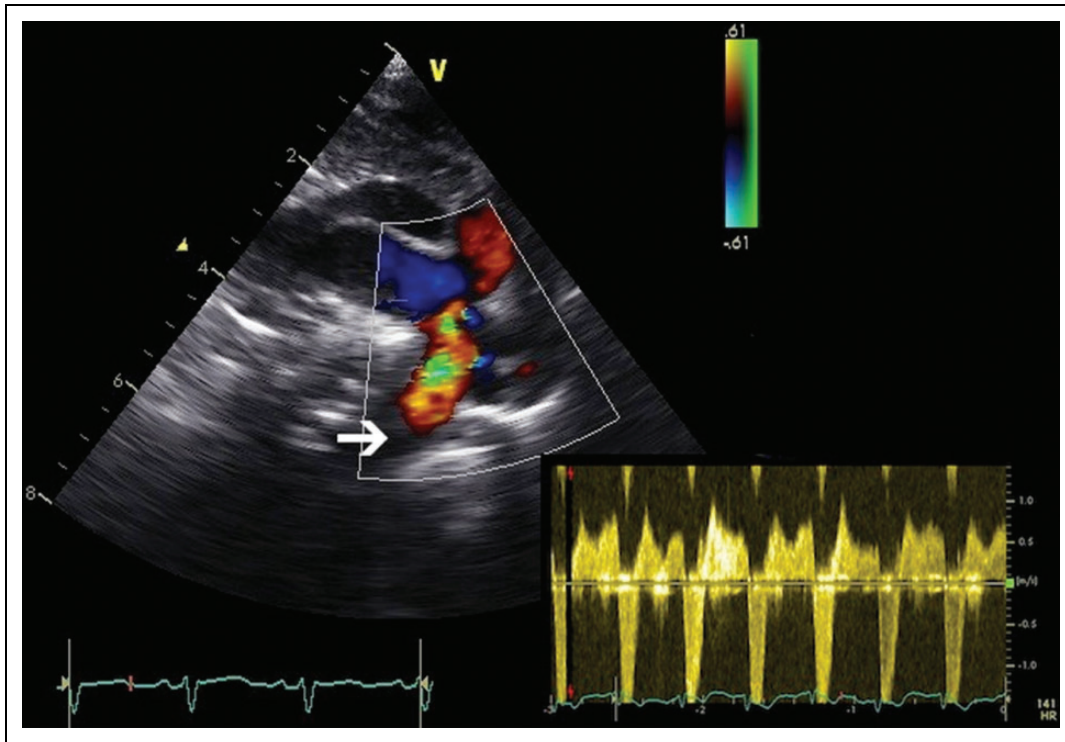
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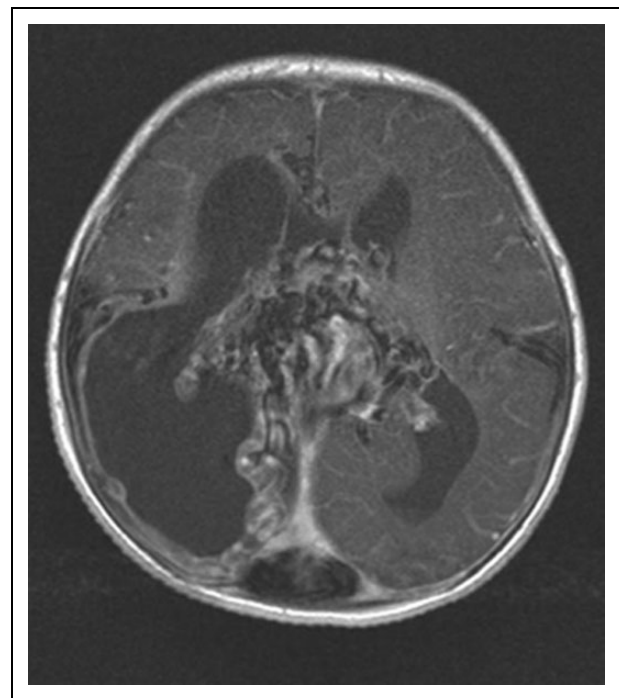
**Figure 1.** Two-dimensional echocardiographic recording of the distal portion of the aortic arch with color flow mapping. Still-frame showing retrograde flow during diastole (in red) caused by diastolic steal toward the cerebral arteriovenous malformation. At the site of the arrow, a pulsed-wave Doppler signal was recorded (inset at the lower right portion of the figure) which documents the holodiastolic retrograde flow in the descending aorta.

suspected because of the diastolic steal phenomenon observed in the descending aorta.

Magnetic resonance imaging showed a vein of Galen aneurysmal malformation, with a maximal diameter of 2.3 cm and ischemic lesions in the left hemisphere and a hydrocephalus (Figure 2). Recurrent seizures were treated with antiepileptic drugs. On day 5 after birth, the electroencephalography showed low voltage with epileptic activity and nearly absent cerebral activity. The option of endovascular treatment, which is the preferred method of treatment in vein of Galen aneurysmal malformation, was discussed. After having discussed the case in the medical team, the authors refrained from endovascular treatment and further intensive care unit treatment because the neurologic prognosis was considered infaust based on the clinical condition and the findings on electroencephalography and magnetic resonance imaging. His parents supported our decision. He died shortly thereafter.

## Discussion

The authors report a case that illustrates the dramatic outcome of a neonate presenting with heart failure and finally being diagnosed with a vein of Galen aneurysmal malformation. The vein of Galen aneurysmal malformation in our patient was of the choroidal type, which can be classified as type I by the classification criteria of Lasjaunias et al.<sup>3</sup>



**Figure 2.** Vein of Galen arteriovenous malformation, magnetic resonance image. Ischemic lesions in the left hemisphere and as a result of the venous congestion, there is a resorption disorder of the cerebrospinal fluid resulting in a hydrocephalus.

This type is the most severe form of the disease, causing high-output cardiac failure in the newborn.<sup>3,4</sup> Furthermore, the symptoms of our patient started at age of less than 5 months, which is also known to be a poor prognostic factor.<sup>5</sup> The unfavorable combination of a Lasjaunias type 1 vein of Galen aneurysmal malformation with severe heart failure and presentation of symptoms at age less than 5 year explains why the clinical course of this patient quickly deteriorated and lead to his death.<sup>5</sup>

In the fetus, high-output cardiac failure may not occur because the low vascular resistance in the cerebral arteriovenous malformations is balanced by the low resistance of the uteroplacental unit. The exclusion of the low resistance placental circulation and the increase in systemic vascular resistance after birth favor flow through the cerebral arteriovenous fistulas, leading to decreased systemic circulation and high output cardiac heart failure.<sup>6</sup>

The size of the fistulas determine the amount of arteriovenous shunting and consequently the time course to the development of cardiac failure.<sup>1</sup> Heart failure develops because of volume overload in the right side of the heart. Due to a low vascular resistance in the head, the majority of left ventricular output is directed toward the head. These 2 mechanisms lead to reduced systemic blood flow, severe lactic acidosis, potentially ischemic multiple organ failure, and persistent pulmonary hypertension of the newborn.<sup>7</sup> During diastole, the systemic perfusion is further reduced by circulatory “steal” to the vein of Galen aneurysmal malformation, which produces the characteristic reversed diastolic flow in the descending aorta. Coronary blood flow is reduced by this diastolic steal phenomenon and combined with increased ventricular pressures can lead to reduced myocardial perfusion or even ischemia, further compromising cardiac function.<sup>6</sup>

Our patient also presented with a hydrocephalus as a result of systemic venous hypertension in the presence of an arteriovenous shunt causing cerebrospinal fluid malabsorption. This cerebrospinal fluid malabsorption is further aggravated by pulmonary hypertension.<sup>5</sup>

Usually, signs of severe heart failure develop shortly after birth. The clinical picture is dominated by findings suggestive of a congenital cardiac disease such as cyanosis, compromised peripheral pulses, and a cardiac murmur. Often cyanosis is present suggestive of persistent pulmonary hypertension of the newborn with signs of right-to-left arterial shunting at the ductal level.<sup>8</sup> A continuous murmur can be heard over the scalp, but this is not routinely checked as our case illustrates.

Chest X-ray imaging may show an enlarged cardiac silhouette with right chamber preponderance, superior mediastinal widening, retrosternal fullness with posterior displacement of the upper trachea, and retropharyngeal soft tissue thickening. The electrocardiography may show ST-T abnormalities. Echocardiography often shows a structural normal heart and great vessels but a dilated and noncompliant right ventricle with signs of suprasystemic pulmonary arterial pressure and right-to-left ductal shunting. The left ventricle on the contrary is often hyperdynamic and not enlarged. Finally, an enlarged

aortic arch and superior systemic veins, especially the superior caval vein, can be observed.<sup>8</sup> Computer tomography and magnetic resonance imaging give a clear demonstration of the intracranial arteriovenous malformation. Cerebral angiography will eventually provide an accurate anatomical definition of all the vessels feeding the fistula. This is essential if treatment is contemplated.

The treatment of vein of Galen aneurysmal malformation involves initial cardiovascular stabilization directed toward improvement in the noncerebral systemic cardiac output. This is achieved by reducing systemic and pulmonary vascular resistance and improving the systemic output and myocardial function of the heart. In most situations, a vasodilator agent, either alone or in combination with low to moderate dose inotropic agents, is needed. Milrinone and inhaled nitric oxide have been found suitable for use in vein of Galen aneurysmal malformation.<sup>1,6,9</sup> Some patients can benefit from treatment with prostaglandin E1. In this way, right-to-left ductal shunt facilitating an adequate systemic circulation is also maintained.<sup>10</sup> However, in about half of patients, cardiovascular stabilization is not achieved.<sup>2</sup>

In the modern treatment of vein of Galen aneurysmal malformation, surgery has little role.<sup>11</sup> Embolization by endovascular treatment is the preferred treatment modality, preferably after the age of 5 to 6 months. A good outcome is to be expected when treatment is performed before significant brain injury has occurred in patients who have been selected carefully.<sup>4,12</sup> A recent systematic meta-analysis of 667 patients revealed that 68% of the patients, including neonates 44%, infants 41%, children and adults 12%, had a good outcome after endovascular embolization.<sup>13</sup> A good outcome was defined as neurologically normal and mild developmental delays. Postembolization mortality was 10% for the total group. Complications such as cerebral hemorrhage, cerebral ischemia, hydrocephalus, leg ischemia, or vessel perforation were seen in 37%.<sup>13</sup> Similar results were found in a review by Khullar et al,<sup>14</sup> which reported 84% good to fair outcome and a 15% mortality rate among 337 patients undergoing endovascular treatment during the time period of 2001 and 2010. A good outcome was defined as neurologically normal or mild developmental delays, and a fair outcome was defined as moderate or severe developmental delays. Neonates showed the worst mortality rate of 36% following endovascular treatment. However, untreated patients with vein of Galen malformation revealed a mortality rate of 77% underlying the great importance of embolization as preferred treatment modality.<sup>14</sup>

In conclusion, vein of Galen aneurysmal malformation presents with signs of congestive heart failure. Vein of Galen aneurysmal malformation is a rare disorder and is often diagnosed late. Considering vein of Galen aneurysmal malformation in the differential diagnosis of neonatal congestive heart failure is important in minimizing diagnostic delay. Initial treatment is targeted at improving noncerebral systemic circulation and reduction in pulmonary vascular resistance. When circulatory stabilization can be achieved, endovascular treatment of the arteriovenous malformation is the preferred

method of intervention. However, despite optimal management, the disorder still carries a high mortality risk.

### Author Contribution

AM is the first author because she wrote the first draft of the manuscript. FH and MS contributed equally and essentially to this work.

### Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

### Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

### Ethical Approval

Written consent was obtained from the parents of the patients to publish information referring to their child.

### References

1. Patel N, Mills JF, Cheung MM, Loughnan PM. Systemic haemodynamics in infants with vein of Galen malformation: assessment and basis for therapy. *J Perinatol.* 2007;27(7):460-463.
2. Berenstein A, Fifi JT, Niimi Y, et al. Vein of Galen malformations in neonates: new management paradigms for improving outcomes. *Neurosurgery.* 2012;70(5):1207-1213;discussion 1213-1204.
3. Lasjaunias P, Terbrugge K, Piske R, Lopez Ibor L, Manelfe C. [Dilatation of the vein of Galen. Anatomoclinical forms and endovascular treatment apropos of 14 cases explored and/or treated between 1983 and 1986]. *Neurochirurgie.* 1987;33(4):315-333.
4. Lasjaunias PL, Chng SM, Sachet M, Alvarez H, Rodesch G, Garcia-Monaco R. The management of vein of Galen aneurysmal malformations. *Neurosurgery.* 2006;59(5 suppl 3):s184-s194;discussion s183-s113.
5. Mortazavi MM, Griessenauer CJ, Foreman P, et al. Vein of Galen aneurysmal malformations: critical analysis of the literature with proposal of a new classification system. *J Neurosurg Pediatr.* 2013;12(3):293-306.
6. Frawley GP, Dargaville PA, Mitchell PJ, Tress BM, Loughnan P. Clinical course and medical management of neonates with severe cardiac failure related to vein of Galen malformation. *Arch Dis Child Fetal Neonatal Ed.* 2002;87(2):f144-f149.
7. Henderson L, Emery DJ, Phillipos EZ, Bhargava R, Olley PM, Lemke RP. Persistent pulmonary hypertension of the newborn presenting as the primary manifestation of intracranial arteriovenous malformation of the Vein of Galen. *Am J Perinatol.* 2000;17(8):405-410.
8. Pellegrino PA, Milanesi O, Saia OS, Carollo C. Congestive heart failure secondary to cerebral arterio-venous fistula. *Childs Nerv Syst.* 1987;3(3):141-144.
9. De Victor D, Le Pommelet C. Medical treatment of congestive cardiac failure from cerebral arteriovenous fistulas at bicetre hospital. *Interv Neuroradiol.* 1996;2(1):49-52.
10. Karam O, da Cruz E, Rimensberger PC. VGAM induced high-flow congestive heart failure responsive to PGE1 infusion. *Int J Cardiol.* 2009;132(2):e60-e62.
11. McSweeney N, Brew S, Bhate S, Cox T, Roebuck DJ, Ganesan V. Management and outcome of vein of Galen malformation. *Arch Dis Child.* 2010;95(11):903-909.
12. Heuchan AM, Bhattacharyya J. Superior vena cava flow and management of neonates with vein of Galen malformation. *Arch Dis Child Fetal Neonatal Ed.* 2012;97(5):f344-f347.
13. Yan J, Wen J, Gopaul R, Zhang CY, Xiao SW. Outcome and complications of endovascular embolization for vein of Galen malformations: a systematic review and meta-analysis. *J Neurosurg.* 2015;123(4):872-890.
14. Khullar D, Andeejani AM, Bulsara KR. Evolution of treatment options for vein of Galen malformations. *J Neurosurg Pediatr.* 2010;6(5):444-451.