

The influence of malformation of Galen vein on the cardiovascular system in a newborn

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Abstract

OBJECTIVES: Asphyxia of the newborn has a varied etiology. Clinical consequences have a broad spectrum of presentations. Arteriovenous malformation associated with an aneurysm of the Galen vein can be the cause of focal ischemic changes in the nervous parenchyma.

RESULTS: The authors report a case of a term newborn (birth weight 4000 grams, Apgar score 7/9). Physical investigation confirmed the presence of a continuous murmur in the area of the anterior fontanelle. Ultrasonic investigation of the brain detected a huge arteriovenous malformation of the Galen vein. Ultrasonic investigation of the heart excluded structural anomaly, but confirmed a huge retrograde flow in the aorta descendens, opened ductus arteriosus with suspected formation of coarctation of the aorta and dilatation of the vena cava superior.

CONCLUSION: Congenital anomaly of the Galen vein has a negative influence on prenatal and postnatal development of the brain of a newborn. In the case of our patient, it led to rapid severe asphyxiated changes of the brain parenchyma. Diagnosis and management were established, yet endovascular therapy was not indicated in the early neonatal period.

INTRODUCTION

The brain of the newborn needs for its good development not only an adequate inflow of oxygen and nutrients but also a functional vessel plexus. Brain arteries supply the brain with oxygenated blood. The cerebral veins are vessels with a thin wall. They are without valves and without muscle layer and are running independently of arteries. Brain veins are divided into superficial and deep ones. The second group comprises the basal vein (*vena basalis*), great cerebral vein (*vena magna cerebri*; *vena Galeni*), lateral direct veins (*vv. directae late-*

rales) and posterior vein of corpus callosum (*v. posterior corporis callosi*) (Holomáňová & Brucknerová 2000; 2003).

The physiological connection between artery and vein is through the arterial plexus (*rete arteriosum*) and venous plexus (*rete venosum*). In the presence of an arteriovenous anomaly, both the arterial and venous plexus are missing in a certain part, presenting a direct pathological connection. This causes a great pressure difference between artery and vein. The higher the pressure differ-

ence, the more expressive are the haemodynamic changes and increased flow. The redirection of blood flow marked as the steal phenomenon is the cause of severe brain ischaemia, presented with disintegration of brain parenchyma. The rapid left-to-right shunting of the blood accelerates and emphasizes the focal ischaemic changes.

In cases of congenital anomaly of vessels, damage of brain functions and brain parenchyma occurs. Arteriovenous brain malformations can be located in the membranes that cover the brain (Suh *et al.* 2005; Gupta & Periakaruppan 2009). Arteriovenous malformation (AVM) of the Galen vein has the highest occurrence rate (Cunliffe 1974; Watson *et al.* 1976; Pellegrino *et al.* 1987). The etiology is unknown. An arteriovenous fistula is the cause of severe congestive heart failure in the newborn, hydrocephalus or seizure activity (Cunliffe 1974; Watson *et al.* 1976; Pellegrino *et al.* 1987). Older children or young adults can present with intracranial haemorrhage (Meyer *et al.* 2000).

CASE REPORT

The family history was without information about congenital anomalies, hypertension, vessel's anomalies. In the third trimester of pregnancy, an arteriovenous malformation was suspected in the brain of the fetus on using magnetic resonance imaging.

The authors present a term newborn (38 gestational weeks, birth weight 4000 grams, birth length 52 cm, Apgar score 7/9). It was the second child from the 3rd pregnancy of healthy parents. The delivery was spontaneous in head position.

At the time of admission (2.5 hour of life), the only pathological finding during physical investigation was the presence of continuous murmur over the anterior fontanelle. The child had no signs of cardiac decompensation, breathing was without ventilator support, without necessity of oxygen inhalation. Ultrasonic investigation of the heart excluded structural anomaly, but confirmed a dilatation of the superior vena cava (8–9 mm), open ductus arteriosus with narrowing on the aortic end (3 mm), a huge retrograde flow from the descending aorta directly to its branches (Figure 1) and formation of coarctation of the aorta with the size of aortic isthmus 6 mm, aorta descendens 3 mm and gradient 20 Torr. Ultrasonic investigation of the brain detected a huge arteriovenous malformation of the Galen vein (size 22 × 25 mm) with many inflow vessels and two huge outflow vessels (Figure 2).

DISCUSSION

An intracranial arteriovenous malformation associated with an aneurysm of the vein of Galen has the most frequent occurrence rate (Pellegrino *et al.* 1987). AVM of the Galen vein has a negative influence on prenatal and postnatal development of the brain of a newborn.

Echocardiography is a method of choice for excluding congenital heart anomaly. It is most important to use a colour Doppler sonography

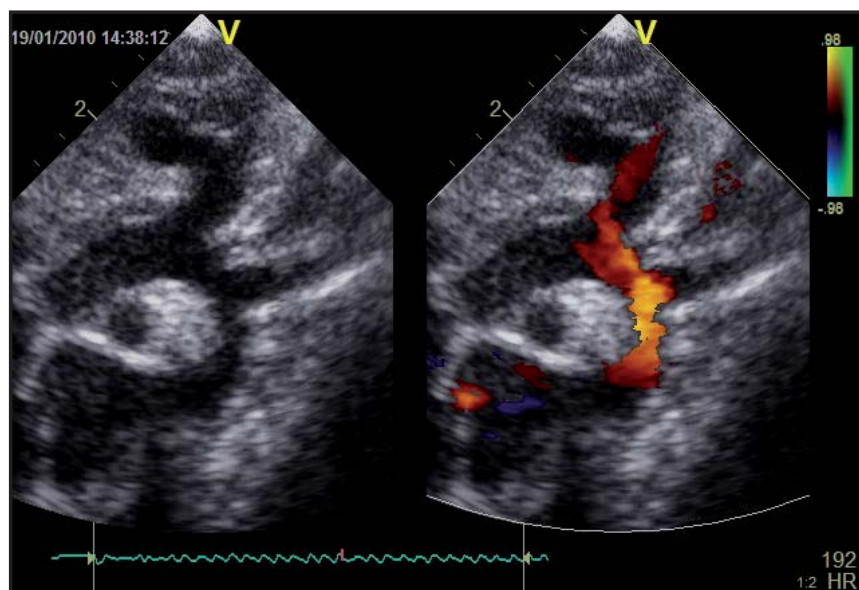


Fig. 1. Ultrasonic investigation of the aorta descendens with confirmation of retrograde blood flow (red colour).

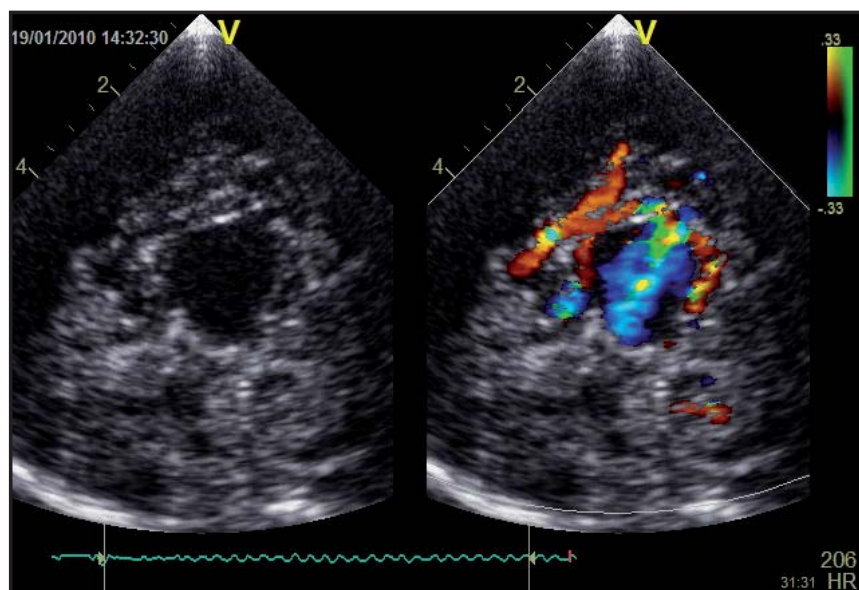


Fig. 2. Ultrasonic investigation of the brain on 1st day of life. Arteriovenous malformation of the Galen vein – an aneurysmal dilatation with many inflow and large outflow vessels with high flow draining into and out.

of the heart and great vessels, including the aortic arch, for confirmation of the pathological direction of blood flow, as performed in the case of our patient. At the site of malformation, the flow was so aggressive that it caused "suction" of the blood from the descending aorta (retrograde flow). The use of echocardiography decreases the need of cardiac catheterization. In comparison with patients described in the literature (Pellegrino *et al.* 1987; Cunliffe 1974), the complication in the case of our patient was not only a change of blood flow but also the danger that severe coarctation of the aorta may develop.

Sonography of the brain is a simple noninvasive method which confirms the presence of congenital vessel anomaly and establishes the size of blood flow through the afferent and efferent vessels. Magnetic resonance imaging with angiography of the brain provides detailed information about congenital anomaly.

Therapy of arteriovenous malformation is symptomatic and surgical. The aim of symptomatic treatment is to stabilize the patient, to obtain normoxemia and physiological values of vital functions. The biggest problem in the case of our patient was the formation of coarctation of the aorta, which called for a continuous infusion of prostaglandins. Congestive heart failure is treated with digoxin and diuretics (Cunliffe 1974).

Due to the developing endovascular techniques, the prognosis of newborns with this congenital anomaly has improved. For endovascular procedures (N-butyl-2-cyanoacrylate; coil) the most important is the size of the arteriovenous anomaly, localization and status of brain parenchyma (Dorfer *et al.* 2009; Saraf *et al.* 2010; Shi *et al.* 2008). In a case of progressive rapid changes of neuronal tissue, endovascular surgery is not indicated.

Mortality is very high in the neonatal period. Antenatal diagnosis can be useful and can improve the outcome. Emergent diagnosis and management is most important and crucial (Doren *et al.* 1995; Has *et al.* 2003).

In our case, the presence of arteriovenous malformation of the Galen vein led to rapid severe atrophy of nerve parenchyma of the brain. The neuronal damage was due to severe focal asphyxia. Confirmation of this congenital anomaly and early management of arteriovenous malformation was done. Due to severe and rapid brain damage (diffuse brain atrophy, dilatation of IIIrd ventricle), endovascular therapy was contraindicated in the case of our patient.

CONCLUSION

Prenatal diagnosis and early postnatal treatment are important in preventing not only heart failure but mainly asphyxiated brain damage, and finally in decreasing postnatal mortality and morbidity. Sometimes the asphyxiation changes in the brain are so rapid that possible therapeutic intervention is contraindicated.

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