

Vein of Galen aneurysm:
Aneurysmal characteristics
and clinical features as
predictive factors

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Congenital vascular anomalies of the central nervous system may cause acute ischemia and hypoxia to the brain, as well as high output cardiac failure. Moreover, intracerebral and subarachnoid hemorrhages can occur at any age. Diagnosis and consideration of therapeutical interventions coupled with a risk assessment in the newborn pose an enormous challenge to clinicians.

Pregnancy was induced by ICSI in a 33-year-old woman and resulted in dichorial diamniotic twins. Because of a suspected cerebral vascular malformation in one of the fetuses on ultrasound exam, the mother was referred to our hospital at 32 0/7 weeks of gestation. Ultrasonography and MRI of the fetal brain revealed an aneurysm of the Galenic vein with beginning heart failure. Because of rapid progressive cardiac decompensation of this twin, a Caesarean section was performed two days later.

Twin B, a girl, was born with Apgar scores of 5, 5 and 5 at 1, 5 and 10 minutes, respectively. The umbilical artery pH was 7.21. Because of respiratory insufficiency and bradycardia, intubation and mechanical ventilation with an FiO₂ of 0.6 were necessary. Blood gas analysis 12 minutes after birth showed a mixed acidosis (pH 7.01, pCO₂ 11.5 kPa, BE -10 mmol/l), with subsequent normalisation to pH 7.36, pCO₂ 5.1 kPa, and a BE -3.4 mmol/l on admission to the NICU at the age of 30 minutes.

INTRODUCTION

CASE REPORT

On physical examination, birth weight, birth length and head circumference were all within the normal range. Cardiac auscultation revealed a 3/6 systolic heart murmur radiating to the axillary space and to the head. The precordium was hyperactive and a pulsating mass in the neck was seen. Apart from hepatomegaly, the remaining of the physical examination was normal (Video 1).

The cerebral ultrasonography showed a dilated left lateral ventricle with a large, strongly perfused cystic structure below the right lateral ventricle, reaching far across the midline (Fig.1). Additionally, bilateral echodense non-homogenous brain parenchymal lesions were seen within the fronto-parietal region. The cardiac ultrasound revealed severe eccentric right ventricular hypertrophy, significant insufficiency of the tricuspid valve and pulmonary hypertension. In addition, there were dilatation of the ascending aorta and of the arteries supplying the brain, backward flow in the descending aorta, reduced cardiac contractility and hypercirculation. On chest X-ray cardiomegaly was found without evidence of any pulmonary pathology (Fig. 2).

In spite of maximal vasoactive and inotropic support, the newborn girl showed rapid progressive signs of cardiac decompensation with arterial hypotension, anuria, centralisation and metabolic acidosis. In a multi-disciplinary assessment with our interventional neuro-radiologists, the situation was considered to

carry a very poor prognosis, and the condition was judged to be inoperable because of the intractable high output cardiac failure. Therefore and after mutual agreement with the parents, intensive care treatment was withdrawn on the second day of life. The girl was extubated and died rapidly in the arms of her parents.

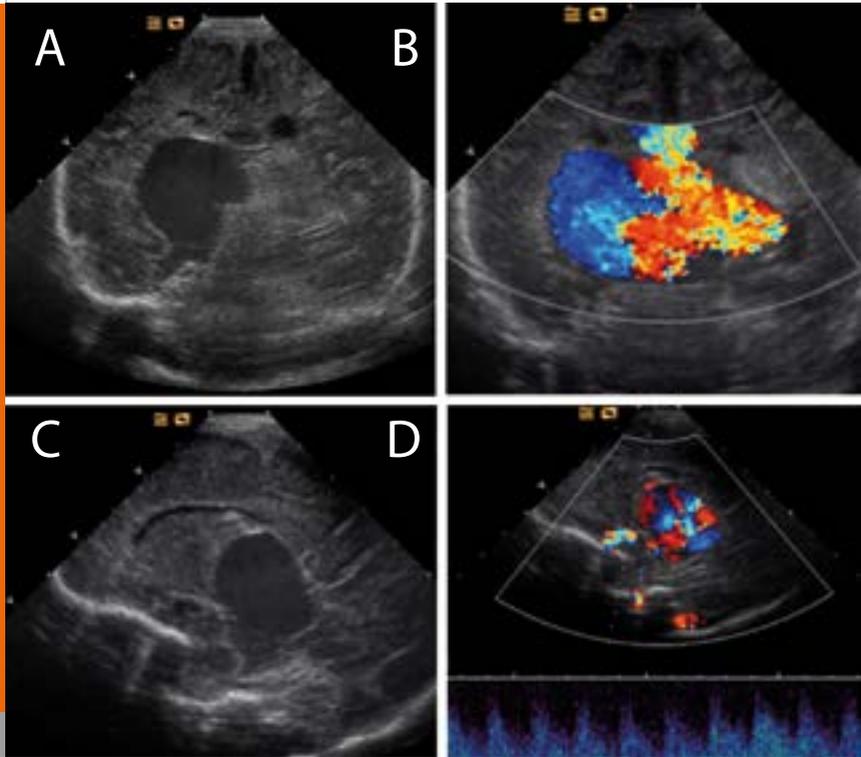


Fig.1

Cerebral ultrasound (A, B: coronal views, C, D: parasagittal views): mildly dilated left ventricle, strongly perfused cystic structure below the right ventricle reaching across the midline.

**Fig.2**

Chest X-ray showing marked cardiomegaly and no evidence of pulmonary pathology.

DISCUSSION

Various forms of arterio-venous aneurysms can be differentiated: congenital (familial), traumatic and mycotic (1). Congenital arterio-venous aneurysms are rare, however of great clinical relevance when present (2). This malformation can lead to a 2- to 4-fold increase of the cerebral perfusion with a considerable risk of hypoxic-ischaemic brain damage (3). With cardiac decompensation due to volume overload, the brain can be further damaged through cerebral hypoperfusion (3). The clinical features have been discussed extensively in the COTM reported by Karam and Rimensberger in September 2008 ("Tachypnea as the first sign of congestive heart failure due to a vein of Galen aneurysmal malformation"). Prognosis depends largely on the extent and localisation of the aneurysm (4). Aneurysms in a median localization and those without manifest heart failure at birth have a better prognosis. Para-median aneurysms spreading laterally across the midline carry a dim prognosis. Signs of heart failure at birth, as in this case, are poor prognostic factors. Para-median vascular aneurysms seem to have a different wall structure which ruptures more easily (5). The prognosis is better in those cases where, after successful conservative treatment of cardiac insufficiency in the neonatal period, the aneurysm either regresses by spontaneous thrombosis, or is removed surgically after the early neonatal period (2, 6). Possible alternative therapeutic options are clipping or intravascular embolization (1, 5). Post-interventional bleeding and hemolysis when the aneurysm is only partially embolized are known risks.

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CONTACT

Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch