Tachypnea as the first sign of congestive heart failure due to a vein of Galen aneurysmal malformation
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Tachypnea is a common feature in newborns. The most frequent etiologies are transient tachypnea of the newborn, respiratory distress syndrome, congenital pneumonia and pneumothorax. More unusual causes are persistent pulmonary hypertension, congenital heart disease or central respiratory drive problems. Tachypnea may be one of the first signs of congestive heart failure. When a newborn presents with signs of congestive heart failure without structural heart disease, an extracardiac high-velocity shunt must be considered.

We present a male infant born by vaginal delivery at 40 weeks of gestation after an uneventful pregnancy. Neonatal adaptation was excellent, with Apgar scores of 8, 9, 10 at 1, 5 and 10 minutes, respectively. At 18 hours of life, brief repetitive movements of his left arm were noted. At 32 hours of life, he developed respiratory distress with tachypnea and perioral cyanosis. At the same time, massive hepatomegaly was noted.

Chest X-ray showed cardiomegaly and increased pulmonary vascular markings (Fig. 1). Echocardiography revealed normal cardiac anatomy with severe global volume overload and suprasystemic pulmonary hypertension with flattening of the interventricular septum, compression of the left ventricle and right-to-left shunting through the foramen ovale. The ductus arteriosus was closed. Cerebral ultrasound examination was indicative for a large vein of Galen aneurysmal mal-
formation (VGAM), which was confirmed by magnetic resonance imaging (Fig. 2 and 3).

The further course was unfavorable with persistent severe suprasystemic pulmonary hypertension and rapidly progressive intractable cardiac failure despite administration of furosemide, continuous positive airway pressure, 100% oxygen and iNO, leading to progressive lactic acidosis (Fig. 4) and eventually to multiple organ failure. Prostaglandin E1 (PGE1) infusion was started on the third day of life, while awaiting percutaneous occlusion of the VGAM. This drug rapidly reversed signs of cardiogenic shock and diminished blood lactate concentrations (Fig. 4).

During percutaneous neuroradiological embolization of one of the aneurysmal arteries (Fig. 5), the VGAM ruptured, leading to massive and fatal intracerebral hemorrhage.

VGAM is a rare (less than 1/25’000 deliveries) (1), but potentially life-threatening cerebral vascular malformation. The VGAM derives from the persistent median prosencephalic vein of Markowski, a transitory venous structure that normally disappears by the 11th week of gestation, resulting in direct arteriovenous communications between the developing arterial network and the median prosencephalic vein.
Babygram showing cardiomegaly.
During intrauterine life, the low resistance vascular bed of the placental circulation competes with the cerebral arteriovenous shunt. Switching off the low resistance placental circulation at birth leads to an increased systemic resistance and a sudden rise of blood flow across the cerebral arteriovenous fistula. This requires a compensatory augmentation of cardiac output to maintain systemic perfusion. The excessive flow through the pulmonary circulation leads to pulmonary hypertension. The high-flow extra-cardiac shunt is responsible for low diastolic pressures. Marked cardiac ischemia results from decreased coronary blood flow during diastole in combination with high-output cardiac failure. In addition, VGAM may cause severe ischemic brain injury because of intracranial steal phenomena and low diastolic pressure with absent or even reversed diastolic cerebral blood flow.

Cardiac failure resulting from VGAM is difficult to treat. While some patients are successfully stabilized with diuretics and dopamine or dobutamine, others may need a combination of β-adrenergic agonists, systemic vasodilators and phosphodiesterase inhibitors. The use of high dose β-adrenergic agents (i.e. dobutamine, dopamine and epinephrine) in this setting can worsen cardiac output by inducing tachycardia and thereby shortening diastolic coronary filling time (1). Improvements in contractility might not be substantial because of right ventricular dilatation and ventricular septal interdependence. Supplemental oxygen and iNO, although decrea-
Cerebral T2-weighted MRI showing the VGAM (asterisk) and neighbouring ischemic lesions (hypointense zones, arrow heads).

Raising right ventricular afterload, may induce a massive increase of left preload with subsequent left ventricular failure (2). PGE1 has been used successfully in cardiac failure due to VGAM. PGE1 has a pulmonary vasodilatory effect that may help to diminish right ventricular afterload. It reestablishes ductal patency allowing right volume unload which improves systemic cardiac output by reducing left ventricular compression. PGE1 has
also systemic vasodilatory effects leading to an improvement of systemic output analogous to the effect of a low resistance placental bed (3, 4). Our patient had refractory cardiac failure, despite 100% oxygen, CPAP and iNO. PGE1 was the only treatment that dramatically improved systemic perfusion and reversed progressive lactic acidosis (Fig. 4).

Advances in interventional neuroradiology have significantly improved the outcome of patients with VGAM. Timing of endovascular interventions is determined by

Angio-MRI showing VGAM with aneurysmal sac (arrow heads) and multiple entering arteries and venous drainage (asterisk).
the degree of hemodynamic instability. After emergency embolization in neonates with intractable hemodynamic instability, the neurological outcome was disastrous despite apparently normal pre-intervention brain imaging (5). Embolization at an age of 5 month has a much better prognosis because of the more mature vascular system. The decision whether an immediate intervention or an initial conservative management is indicated can be based on a 21-point scale presented by Lasjaunias and co-workers, which takes in account the function of several organ systems (6) (Tab. 1). Patients who are hemodynamically stable have a better prognosis if the VGAM is treated beyond the neonatal period, whereas patients who are hemodynamically compromised must undergo urgent endovascular
intervention. VGAM related cardiac failure presenting in the neonatal period with multiple organ failure is associated with a high rate of cerebral infarction and thus a poor prognosis for long-term neurodevelopmental outcome. Therefore, improving and maintaining stable cardiac function is essential in order to perform an endovascular embolization as late as possible. Urgent embolization aims only at partial occlusion of the AV shunt (30-50%) to control its cardiovascular impact. Residual vascular connections are addressed when the patient is older and his vascular system is more mature (5).

*Catheter intervention showing the arterial supply (a) and venous drainage (b) of the VGAM.*

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<th>Score</th>
<th>Cardiac</th>
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<th>Respiratory</th>
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<td>Normal</td>
<td>Normal</td>
<td></td>
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<td>Subclinical EEG anomalies</td>
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<td>Non-convulsive CNS signs</td>
<td>Severe tachypnea</td>
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<td>Isolated convulsion</td>
<td>Assisted ventilation, FiO2 &lt; 25%</td>
<td>Hepatomegaly, normal function</td>
<td>Transient anuria</td>
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<td>Assisted ventilation, FiO2 &gt; 25%</td>
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<td>Assisted ventilation</td>
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