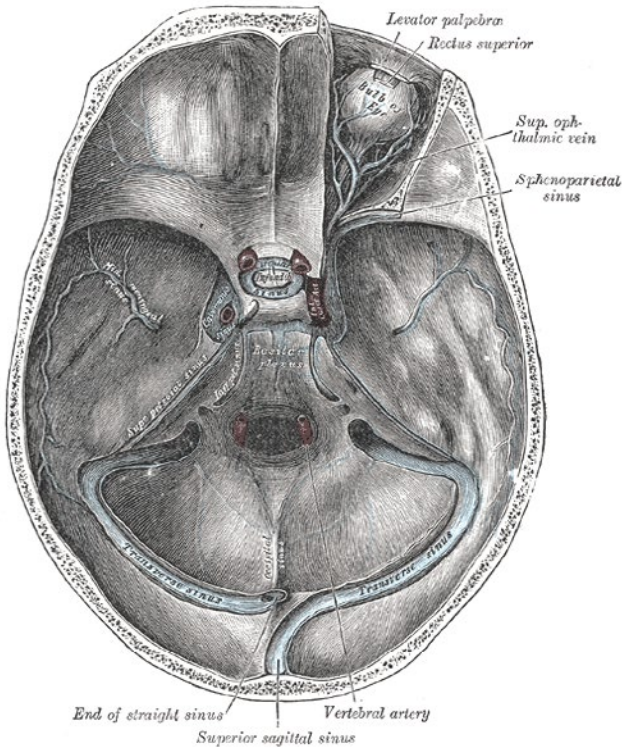
SWISS SOCIETY OF NEONATOLOGY

When a murmur does not come from the heart

February 2020



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Sinus transversii (source: www.de.wikipedia.org)

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INTRODUCTION

Cerebral vascular arteriovenous malformations are rare but potentially life threatening. There are three major types of cerebral vascular arteriovenous shunts in neonates: 1) vein of Galen aneurysmal malformation, 2) pial arteriovenous malformation and 3) dural sinus malformation (1). Diagnosis is not always possible on prenatal imaging. Postnatally, subtle signs of cardiac failure and a murmur heard over the fontanelle can lead to early diagnosis and early treatment. After birth, these malformations can rapidly lead to high-output cardiac failure when left untreated. Later presentations include macrocephaly, seizures or mental retardation.

Transarterial endovascular embolization is the treatment of choice, especially in emergency situations when heart failure cannot be controlled with medications. We report the case of a newborn infant with unexplained respiratory distress at birth, rapidly developing cardiac failure due to a high-flow dural sinus malformation.

CASE REPORT

This baby girl was born at 38 weeks of gestation by Cesarean section (because of maternal fatigue) after an uneventful pregnancy. Birth weight was 3100 g (P10 – 25), birth length 49 cm (P25) and head circumference 36 cm (P75 – 90). She immediately showed signs of severe respiratory distress for which she was put under manual CPAP. The initial chest X-ray was felt to be consistent with retained fetal lung fluid, but also showed slight cardiomegaly (Fig. 1). Due to her respiratory distress, the baby was transferred to a neonatal level III unit.

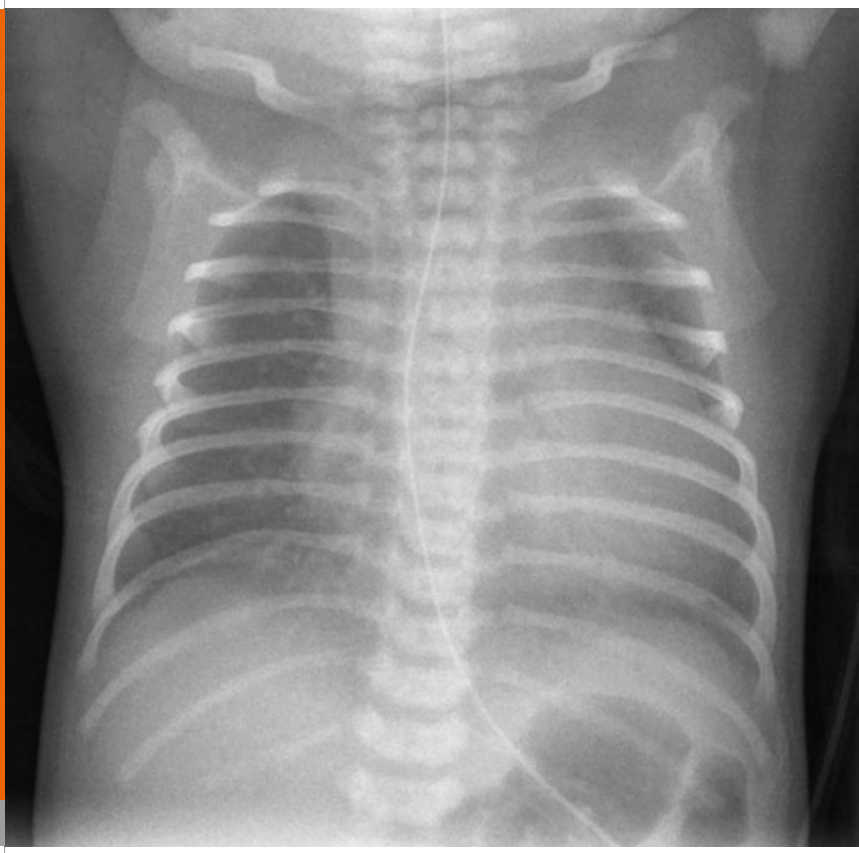

Fig. 1

Chest X-ray on DOL 1: findings consistent with retained fetal lung fluid, mild cardiomegaly.

CPAP was stopped after 3 days, but she remained tachypneic with failure to thrive. Echocardiography showed tricuspid regurgitation, which was interpreted as a minor Ebstein's anomaly. High Flow Nasal Canula (HFNC) was started on DOL 7 due to worsening of respiratory distress.

On DOL 9, her condition suddenly deteriorated with signs of cardiogenic shock. Echocardiography now showed supra-systemic pulmonary hypertension without structural abnormalities. On clinical examination, a murmur was heard over the anterior fontanel and rapid growth of the head circumference was noted. Cerebral ultrasound showed abnormal Doppler flow signals in the left occipital lobe (Fig. 2). She was intubated and iNO was started. MRI on DOL 10 revealed a high-flow arteriovenous malformation located in the left transverse sinus explaining the patient's high-output cardiac failure (Fig. 3 – 5).

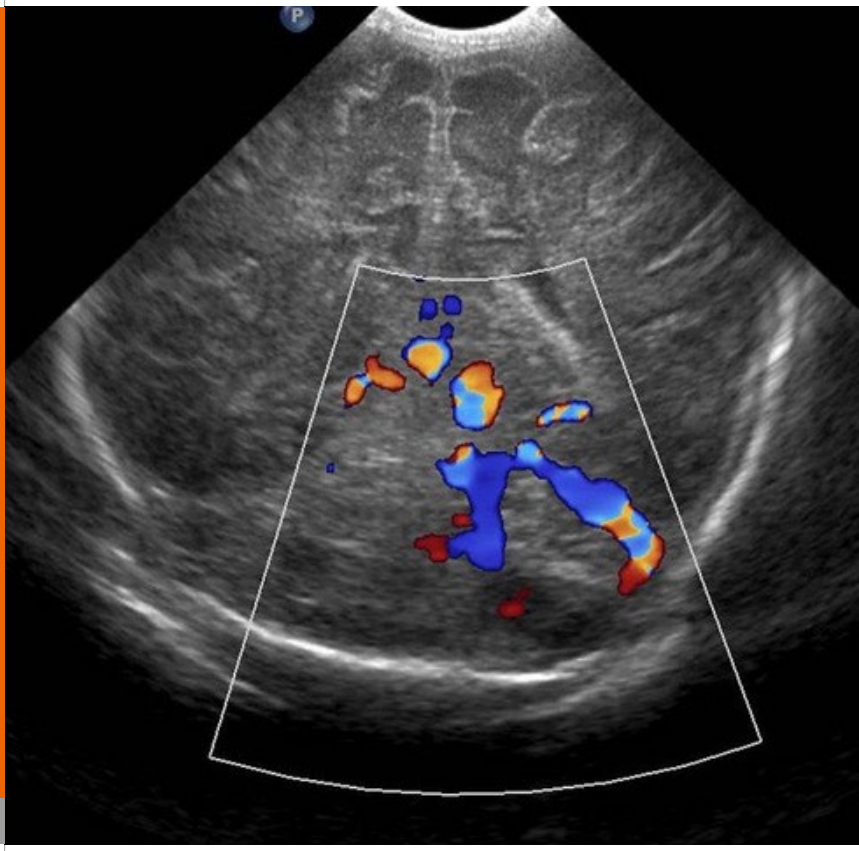

Fig. 2

Cranial ultrasound examination on DOL 9:
increased Doppler flow signal in the posterior fossa.

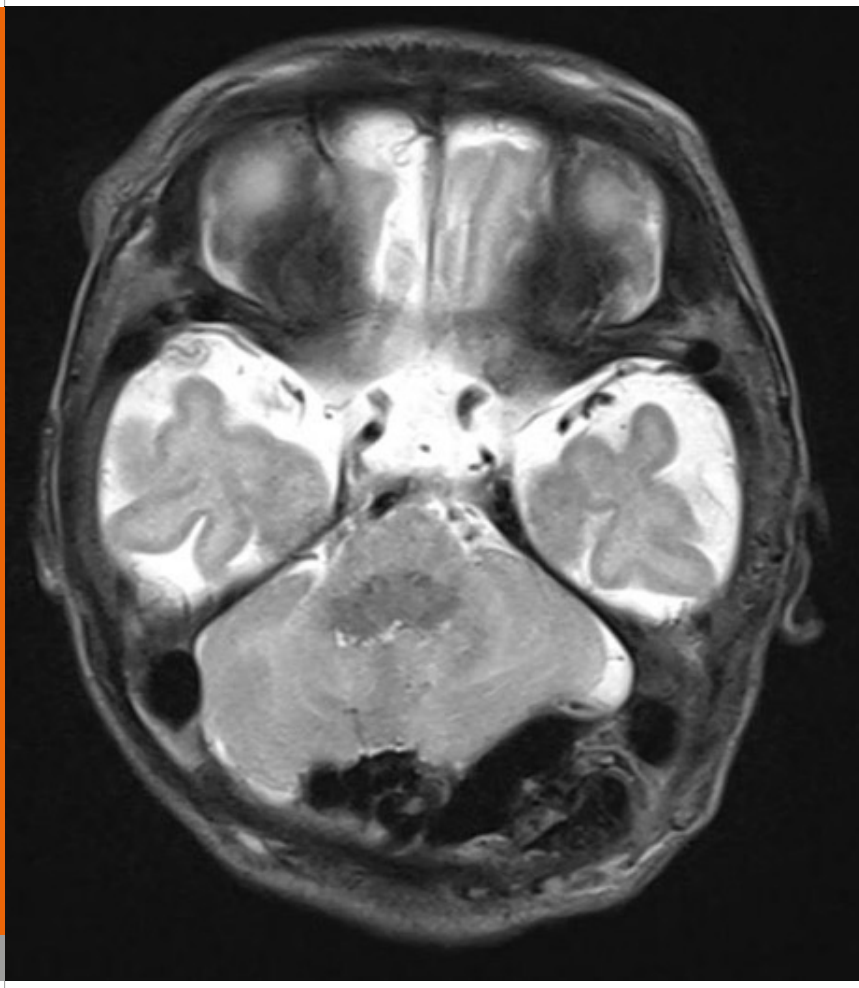

Fig. 3

MRI examination on DOL 10 (T2-weighted images): high-flow arteriovenous malformation located in the left transverse sinus, extending into the posterior fossa (note flow void of the vascular structures).

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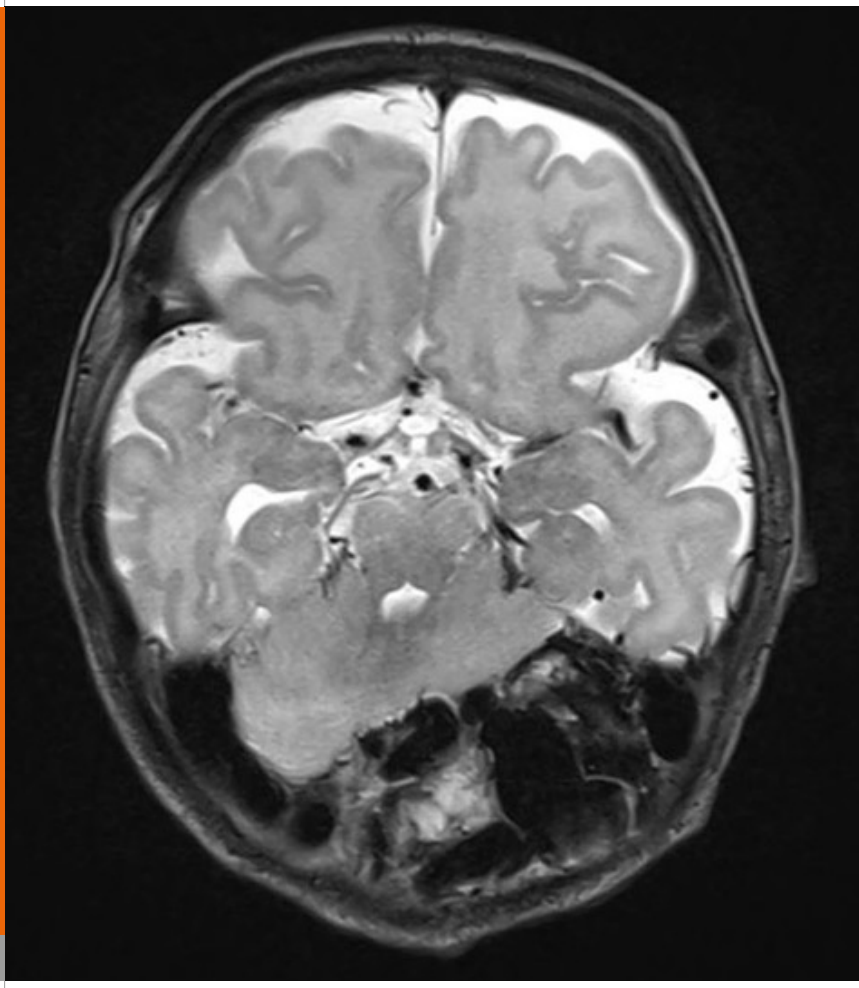

Fig. 4

MRI examination on DOL 10 (T2-weighted images): high-flow arteriovenous malformation located in the left transverse sinus, extending into the posterior fossa (note flow void of the vascular structures).

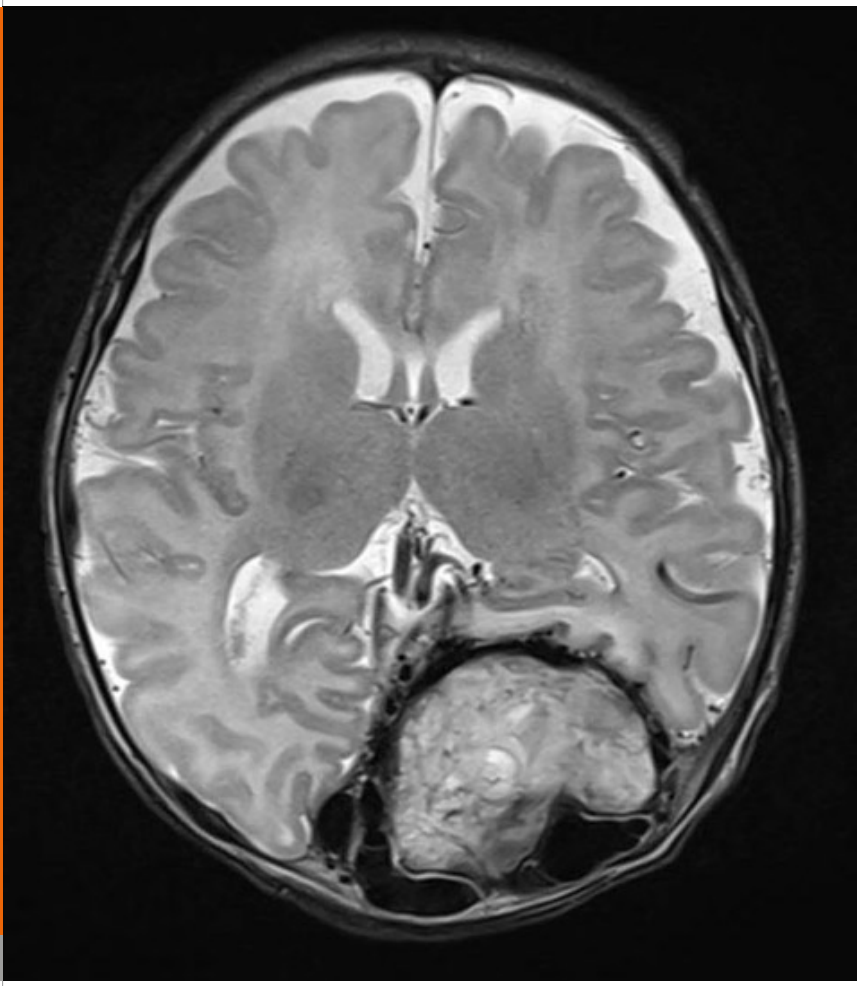

Fig. 5

MRI examination on DOL 10 (T2-weighted images): high-flow arteriovenous malformation located in the left transverse sinus, extending into the posterior fossa (note flow void of the vascular structures).

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On DOL 12, embolization of the vascular malformation was performed with transarterial histoacryl injection after catheterization of the right femoral artery (Fig. 6) as previously published (2). About 60 % of the shunt was successfully occluded.

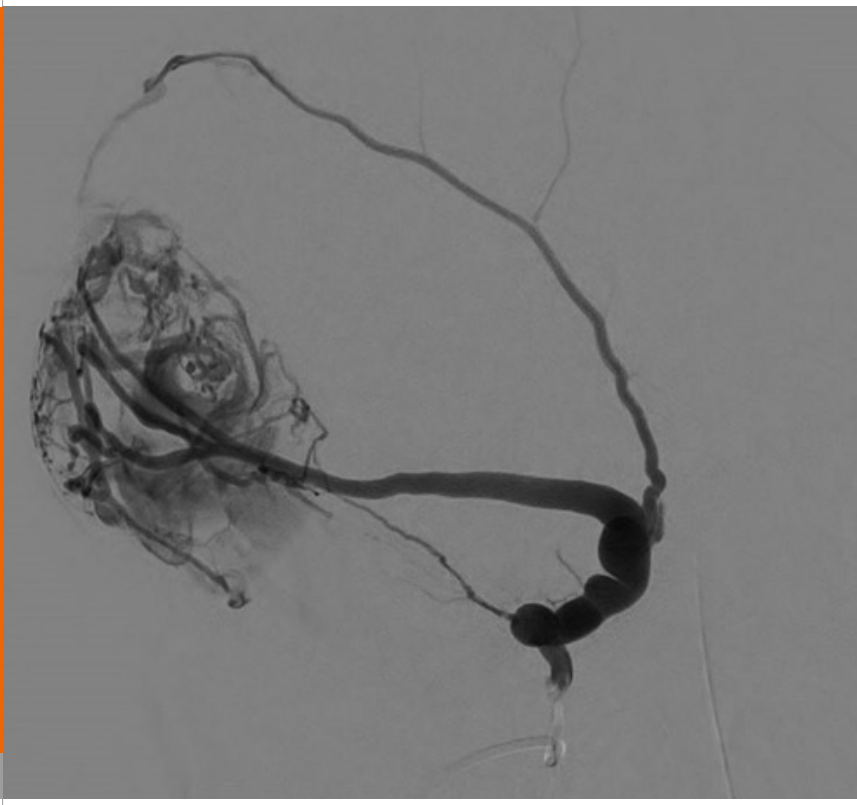

Fig. 6

Angiography on DOL 12: demonstration of a large arteriovenous malformation prior to The first embolization procedure.

Postintervention, she was started on diuretics and anticoagulated (at a curative dose) to prevent excessive clotting of the malformation (3). She was extubated on DOL 15 but had to be reintubated three days later because of worsening signs of cardiac failure. A second embolization procedure was performed on DOL 19 via the left femoral artery, and about 90 % of the initial shunt was occluded. She was extubated to nasal CPAP on DOL 26. Subcutaneous enoxaparin at an initial dose of 1.5 mg / kg twice a day (aiming for an anti-Xa activity between 0.5 and 1) was started to prevent any extensive thrombosis and help to obtain good venous remodeling.

Her condition improved slowly, and nasal CPAP could be discontinued on DOL 54. Neurological examination at two months of life was unremarkable. Cerebral MRI documented important reduction of the dural sinus malformation. The patient was finally discharged home on DOL 68 with enoxaparin 1.5 mg / kg s.c. twice a day.

At 4 months of life, clinical examination was normal, and no murmur was heard over the anterior fontanel. An MRI confirmed treatment success with adequate vascular remodeling. Parenchymal damage appeared to be limited and unchanged to previous MRIs; therefore, the baby is expected to have a good prognosis. Enoxaparin was stopped on DOL 119.

DISCUSSION

This case report highlights the importance of a careful exam after birth, especially in patients presenting with unexplained respiratory distress and evidence of cardiac failure. Auscultation over the anterior fontanel can provide an important clue if a murmur is heard. This clinical finding is highly suggestive of an intracranial high-flow arteriovenous shunt leading to high-output cardiac failure.

In the neonate, the most frequent cerebral arteriovenous malformation involves the vein of Galen. In fact, the vein of Markowsky (5), an embryonic precursor of the vein of Galen located on the choroidal network of the third ventricle, is involved. Pial fistula are located under the pia mater at the surface of the cortex. The least frequent arteriovenous malformations are dural sinus malformations located in the dura (4).

Dural sinus malformations can be diagnosed during the antenatal period. On fetal ultrasound, dilatation of a sinus (also called "giant lake") may be seen. In other patients, it is discovered when they are evaluated for hydrops fetalis due to severe congestive heart failure.

More commonly, however, diagnosis is made after birth in patients with severe congestive heart failure. Increased venous return from the superior vena cava leads to volume overload of the right ventricle already

during the fetal period. After birth, when pulmonary blood flow increases, volume overload occurs in all four chambers and leads to persistent pulmonary hypertension with right to left shunt over the patent ductus arteriosus and the foramen ovale. Closure of the ductus arteriosus leads to significant right-sided heart failure and sometimes to sub-myocardial infarction (6). In our patient, a minor form of Ebstein's anomaly was suspected when tricuspid regurgitation was seen. In retrospect, this was due to developing congestive heart failure due to pulmonary overload. During infancy, hydrodynamic disorders can occur due to increased cerebral venous pressure caused by the shunt, which may lead to hydrocephalus, brain damage and psychomotor delay (7). In addition, when extensive venous thrombosis develops, platelet consumption can occur and lead to intracranial hemorrhage.

Neuroimaging is necessary to better characterize the lesions. Cranial ultrasound is widely available, can be performed at the bedside, and sometimes allows diagnosis, especially when color Doppler is used. However, its sensitivity is influenced by the experience of the investigator and limited by the acoustic shadow of the skull; in addition, lesions in the posterior fossa may be hard to visualize when not using the mastoid window. Cerebral CT with contrast enhancement is easy to perform, but brain MRI is the imaging study of choice to better characterize the type of malformation

and to identify parenchymal injury. Arteriography is not required: in fact, access sites should be preserved for later endovascular treatment (8).

Because of its low invasiveness and high efficiency, transarterial embolization with glue (N-butyl-cyanoacrylate) via a femoral artery is the treatment of choice (9 – 10). Timing of the intervention depends on the type of lesion and the clinical course. Surgical treatment has very limited indications (some pial arteriovenous malformations or pial arteriovenous fistula) and is associated with high morbidity and mortality rates.

Anticoagulation is an important adjunct and facilitates venous remodeling (3). In a recent report, the effects of curative doses of low molecular weight heparin given after embolization procedures were described. The intervention appeared to be safe and effective to treat extensive venous thrombosis or to prevent thrombosis until satisfactory vascular remodeling was obtained.

See also: COTM 09/2008:
Tachypnea as the first sign of congestive heart failure due to a vein of Galen aneurysmal malformation

See also: COTM 04/2010:
Vein of Galen aneurysm: Aneurysmal characteristics and clinical features as predictive factors

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