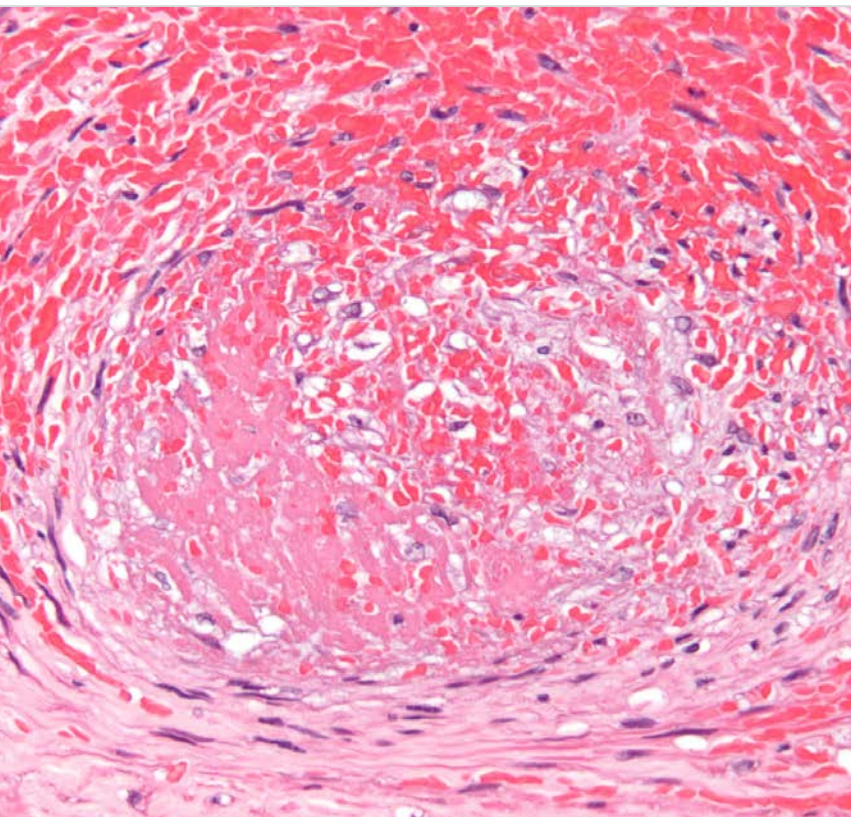
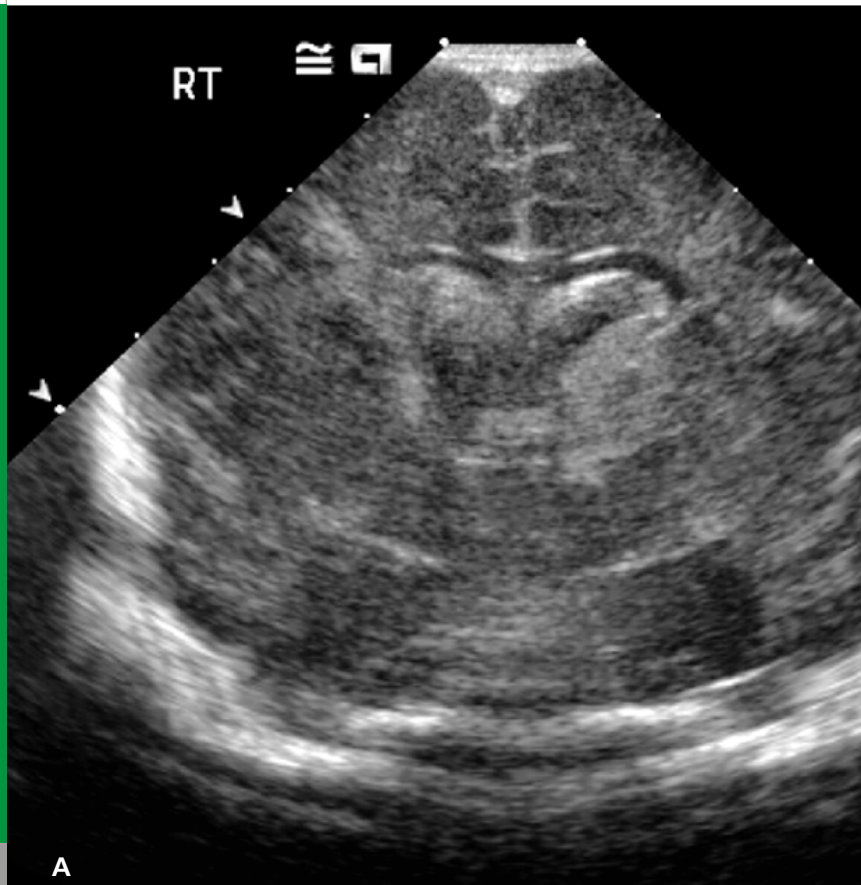


Cerebral venous sinus
thrombosis in a preterm
infant: associated brain
injuries

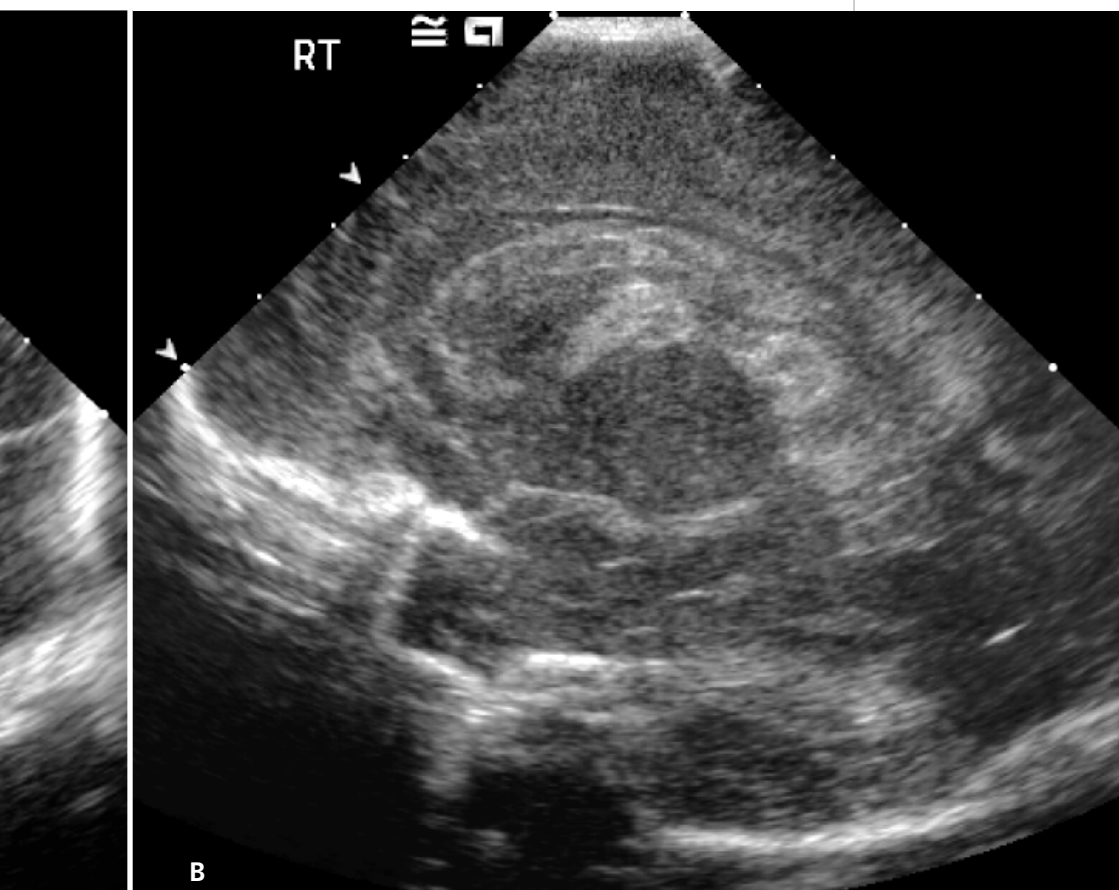
November 2014



Cerebral venous sinus thrombosis (CVST) is rare in the pediatric population. The incidence in Switzerland is 0.56/100'000 in children below the age of 16 years (1). However, the number of cases might be underestimated because of nonspecific clinical presentation and variable sensitivity of different diagnostic methods. Among all children, newborn infants represent the most commonly affected group (1): about half of CVST in children occur during the neonatal period with males being more often affected than female infants (2, 1). In August 2006, a case report on CVST was published on the SGN website, with the focus on the clinical presentation, risk factors and outcome. In contrast to the previously published case, we focus on the pattern of associated brain lesions in preterm infants with CVST.

**Fig. 1****A**

Coronal (A) and parasagittal (B) ultrasound images on day of life 10: bilateral intraventricular hemorrhage and hemorrhage in the left caudate and thalamus; abnormal white matter with loss of differentiation between cortical grey matter and white matter.



CASE REPORT

This female infant was born to a healthy 35-year-old G1/P1 by Caesarean section because of pathological CTG at 34 3/7 weeks of gestation. The pregnancy was complicated by high blood pressure, and oligohydramnios and growth retardation noted at 32 weeks of gestation. Postnatal adaptation was normal with Apgar scores of 8, 9 and 9 at 1, 5, and 10 minutes, respectively, and venous cord pH was 7.45. Birth weight was 1590 g (P3-5), head circumference 29.5 cm (P10-25) and length 43 cm (P10-25). The infant was admitted to the neonatal unit due to prematurity.

The first ten postnatal days were uneventful apart from initial hypoglycemia and unconjugated hyperbilirubinemia which was treated with phototherapy. Routine cranial ultrasound performed on the 4th postnatal day was normal.

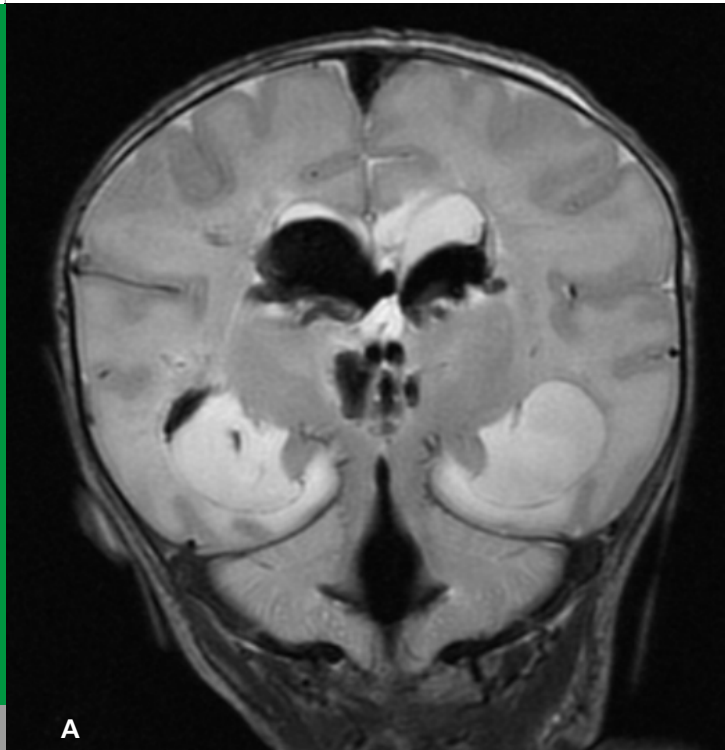
On the eleventh postnatal day, the infant presented with feeding difficulties, pallor, increased muscular tone, opisthotonus and ocular deviation. Following a septic work-up, including a lumbar puncture, which revealed blood stained CSF, antibiotic treatment was started. During the following hour, the child deteriorated rapidly and had to be intubated due to respiratory insufficiency. The infant developed status epilepticus which persisted despite the administration of multiple antiepileptic drugs (phenobarbital, midazolam, levetiracetam). Lactate was increased during the seizures

and a drop in hematocrit from 51% to 28% was noted, hence a blood transfusion was given.

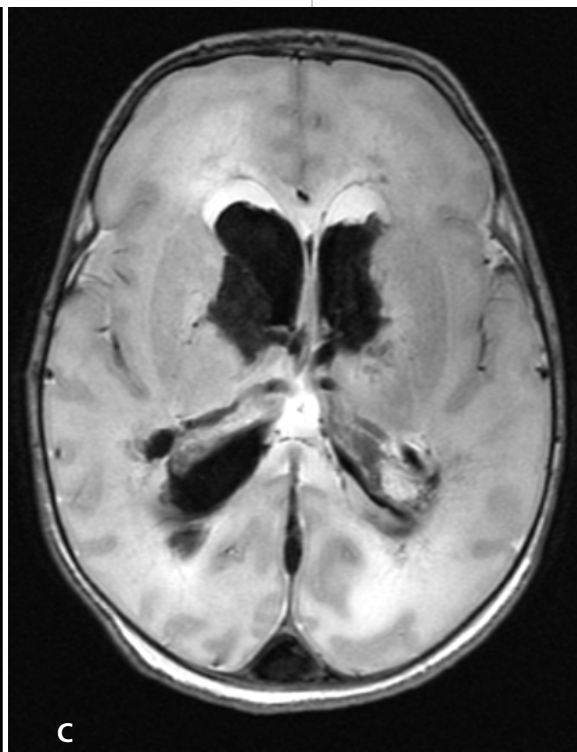
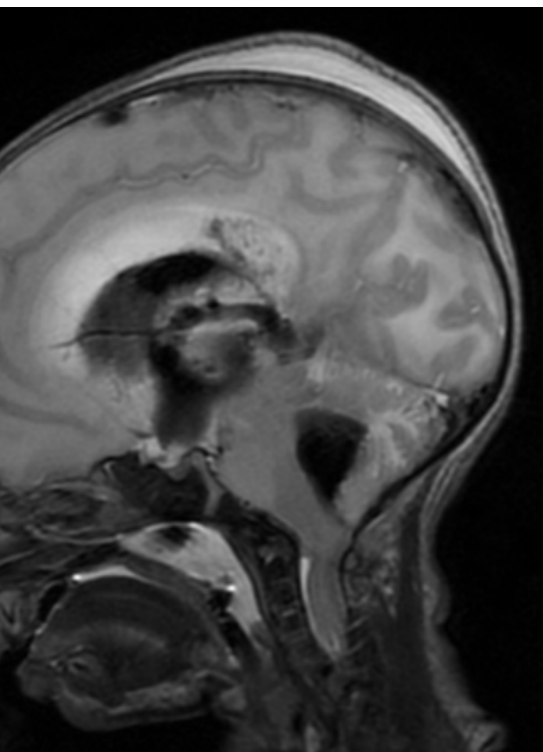
On subsequent cranial ultrasound, large bilateral intraventricular hemorrhages were seen as well as a hemorrhage in the left thalamus (Fig. 1). Furthermore increased white matter echogenicity was seen in frontal and parietal lobes. CVST was suspected at this time and confirmed by magnetic resonance imaging (MRI) on the following day.

MRI showed extensive venous sinus thrombosis in the superior sagittal sinus, the straight sinus, the transverse sinus, the vein of Galen, the internal cerebral veins bilaterally, as well as parietal cortical veins. There were bilateral intraventricular haemorrhages as well as hemorrhages in the basal ganglia and thalamus. In addition, extensive white matter infarction was seen bilaterally with beginning herniation of the medulla oblongata (Fig. 2). Coagulation parameters were normal, and there were no signs of thrombophilia.

Because of the combination of the status epilepticus refractory to treatment, clinical signs of decerebration and the MR findings, the prognosis was thought to be extremely poor. Therefore, it was decided to redirect care, and the girl died shortly afterwards in the arms of her parents.

**Fig. 2**

Axial (A), midline sagittal (B) and coronal (C) T2-weighted MR images: intraventricular hemorrhage, bilateral hemorrhages in the caudate nuclei and thalami and widespread increased signal intensity in the white matter with loss of cortical differentiation consistent with global infarction of white and grey matter; extensive thrombus in the superior sagittal sinus; in addition, there is herniation of the medulla oblongata seen (B).

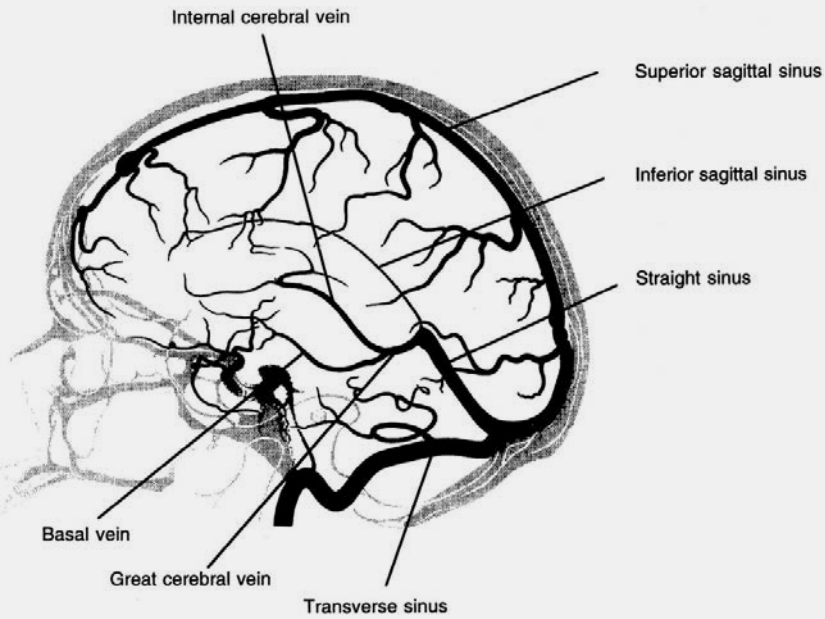


DISCUSSION

The symptoms of CVST in newborn infants are non-specific. The most common symptoms are focal or generalized seizures. When intraventricular hemorrhages and thalamic haemorrhages are demonstrated after the first week of life, a CVST should be suspected and actively sought. Absent or decreased flow in the affected sinus can be detected by Doppler ultrasound examination in approximately half of the infants with CVST, however, thrombosis in the deep venous system might be missed (1).

MRI and/or magnetic resonance venography used for the evaluation of CVST has a sensitivity of 90% and is therefore recommended (3). There are many different appearances of the thrombus on MRI depending on the age of the thrombus. Several pulse sequences need to be performed: T1-weighted, contrast enhanced T1-weighted, T2-weighted, fluid-attenuated inversion recovery, susceptibility-weighted and diffusion weighted sequences, and MR venography (4).

The most commonly affected sinuses in newborn infants are the straight sinus (85%) and the superior sagittal sinus (65%). In the majority of cases, several sinuses are involved. Involvement of the transverse sinus is often seen in combination with that of other sinuses (5). Almost in all cases of CVST associated lesions are found. The associated lesions depend on gestational age with distinctive findings in preterm and in term in-

**Fig. 3**

Cerebral venous sinuses (reproduced from reference 7).

fants, probably related to the vulnerability of the brain during different stages of maturation (5).

In preterm infants, severe white matter lesions affecting the entire periventricular region together with intraventricular haemorrhage(s) are typically seen (2, 5). Other associated brain lesions consist of unilateral thalamic hemorrhage, periventricular hemorrhage, occipital/parietal-occipital infarction or ischemic basal ganglia lesions (5). This is consistent with the findings of our case.

In term infants, the typical finding is intraventricular hemorrhage with unilateral thalamic hemorrhage. Punctate white matter lesions are common in this age group as well; however, these lesions are usually in the frontal region and less extensive than those seen in preterm infants. Occipital/parietal-occipital infarction, infarction of the basal ganglia or periventricular hemorrhage can occur (5).

After recanalization and restoration of blood-flow, a reduction of the extent of the lesions is usually seen in survivors. Late findings are frontal atrophy, cavitation at the site of the previous lesion, delay in myelination and early gliotic changes.

Mortality in newborn infants with CVST varies between 2% and 19% (2, 6). Neurodevelopmental outcome depends on the extent and localisation of the

associated brain lesions and is poor in 30% to 80% of patients. Outcome is particularly poor among preterm infants with CVST because of the associated white matter lesions. Neurodevelopmental impairment consists of sensorimotor deficits, receptive or expressive language problems as well as cognitive and behavioural impairment (6). Thalamic hemorrhage can result in neurobehavioral problems, sensorimotor deficits or visual deficits like strabismus or uncoordinated eye movements. Intraventricular hemorrhage can lead to posthemorrhagic hydrocephalus which might require a ventricular peritoneal shunt. Epilepsy was documented in 19% of survivors (6).

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