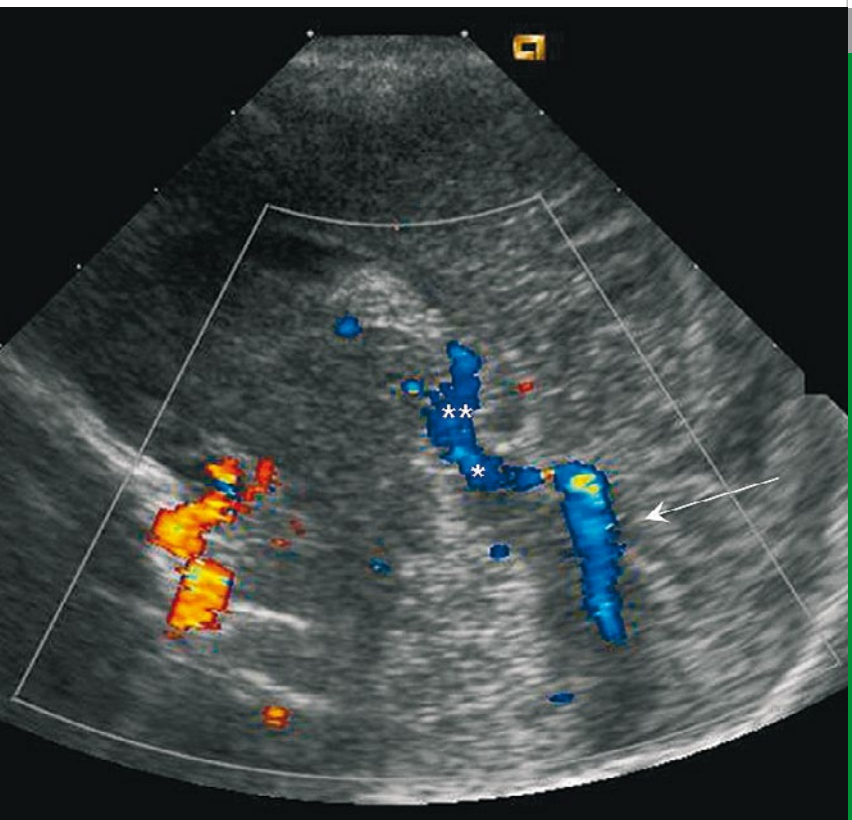
SWISS SOCIETY OF NEONATOLOGY

Neonatal sinovenous thrombosis



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Hauri-Hohl A, Zeilinger G, Pasquier S, Children's Hospital of Aarau, Switzerland

A twin boy (twin A) was born by spontaneous vaginal delivery to a 32-year-old mother at 37 4/7 weeks of gestation. The bichorionic twin pregnancy was complicated by nicotine abuse of the mother (1 pack of cigarettes per day), cervical insufficency requiring placement of a cervical cerclage, tocolysis and lung maturation during the 29th week of pregnancy due to impending premature birth. The infant was admitted to the neonatal unit for 10 days because of intrauterine growth restriction (weight 2080 g, length 43.5 cm, head circumference 31 cm), neonatal hypoglycemia and neonatal thrombocytopenia (platelets of 21 G/l). The number of thrombocytes nomalized (platelets 221 G/l) after transfusion of one unit of platelets and two doses of IVIG. Blood tests confirmed an alloimmune thrombocytopenia.

The boy was admitted to our hospital on the 21st day of life because of refusal to eat, scant diarrhea and continuous crying without obvious cause. On admission he was afebrile and did not show signs of dehydration (normal fontanels) or bleeding. A few hours after admission, his condition suddenly deteriorated with the development of a downward gaze, opisthotonus and bulging of the fontanel. Ultrasonography of the brain was performed promptly and demonstrated intraventricular bleeding and hemorrhagic infarction of the thalamus and pallidum including the internal capsule (Fig. 1-3). A sinovenous thrombosis (SVThr) with complete occlusion of the sinus rectus and partial thromboses of the su-

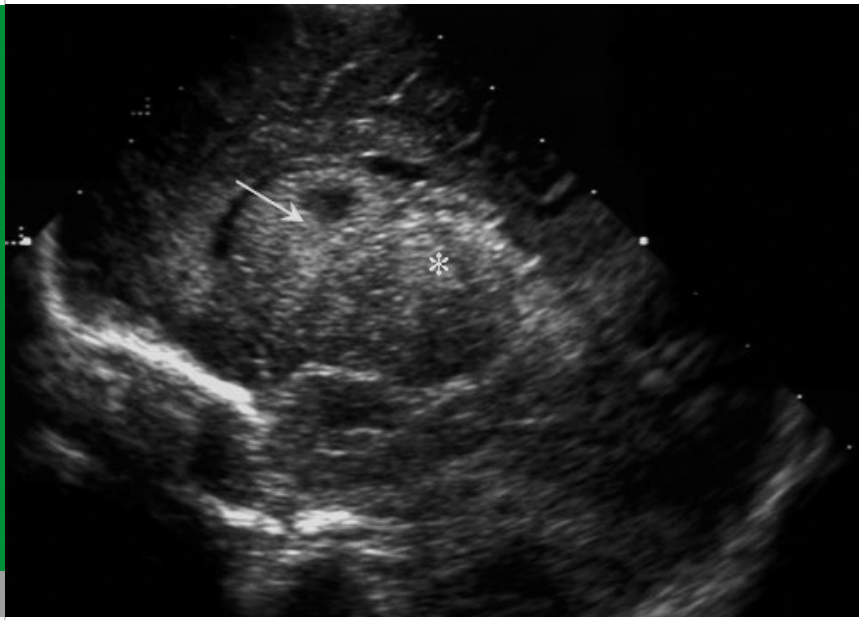


Fig. 1

Cerebral US, parasagittal view: intraventricular hemorrhage (arrow) hemorrhagic infarction of thalamus and basalganglia (*).

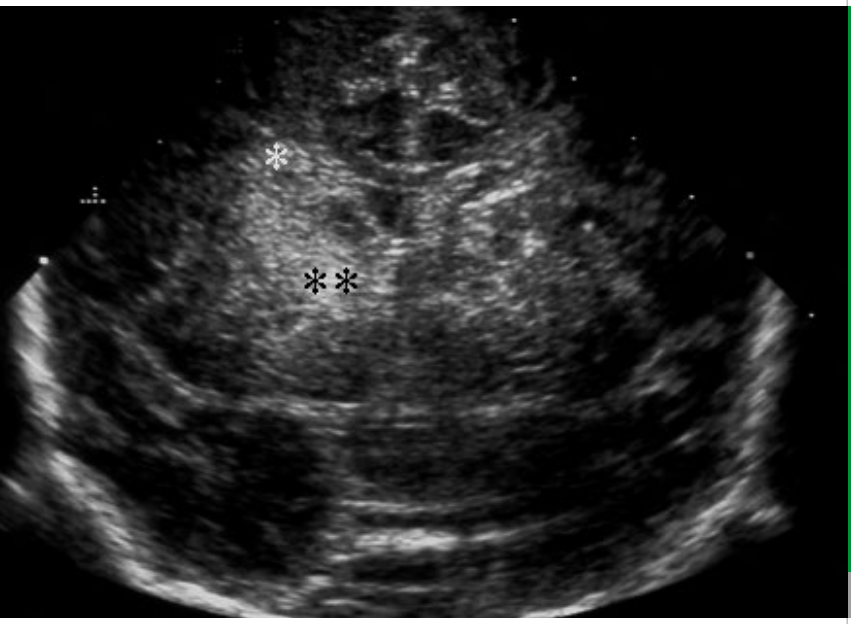


Fig. 2

Cerebral US, coronal view: IVH, hemorrhagic periventricular infarctions (*) and hemorrhagic infarctions of halamus and basal ganglia (**).

perior sagittal and both transversal sinuses were confirmed by MRI (Fig. 4, 5). Anticonvulsive therapy was initiated on the same day due to generalized seizures. The patient did not receive any anticoagulant therapy.

Evaluation for a hypercoagulable state in this child included platelet count, prothrombin time, partial thromboplastin time, fibrinogen level, factor II level, antithrombin III level, functional protein C and S concentrations, APC resistance, functional heparin-co-factor, prothrombin mutation (nt20210 G‡A), factor V mutation (FV:R506Q), anticardiolipin antibodies, and antiglycoprotein antibodies. These investigations revealed a heterozygous factor V mutation.

The boy is now 4 years old and suffers from severe cerebral palsy with affection of speech, vision and motor system. Due to seizures, he remains on anticonvulsant therapy.

DISCUSSION

DeVeber et al. (1) analyzed the risk factors and outcome of SVThr in 160 Canadian children age 0-18 years and compared neonates and non-neonatal children. The indicence of SVThr was 0.67 cases per 100'000 children per year with neonates being most commonly affected (43% of all patients). A similar difference in incidence between neonates and older children was observed in Germany and described by Heller et al (2). The authors reported a yearly incidence of SVThr of 2.6/100'000 neonates compared to 0.35/100'000

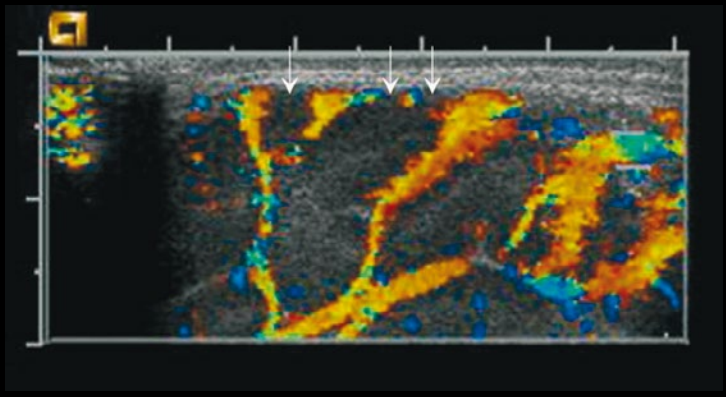


Fig. 3

Cerebral color Doppler study: thrombosis of the superior sagittal sinus (for comparison, see Fig. 7).

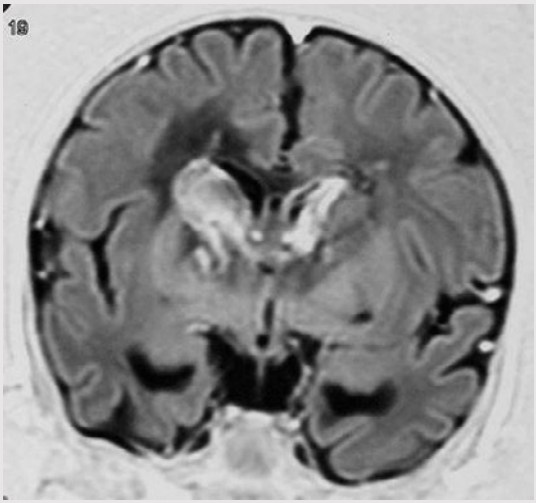
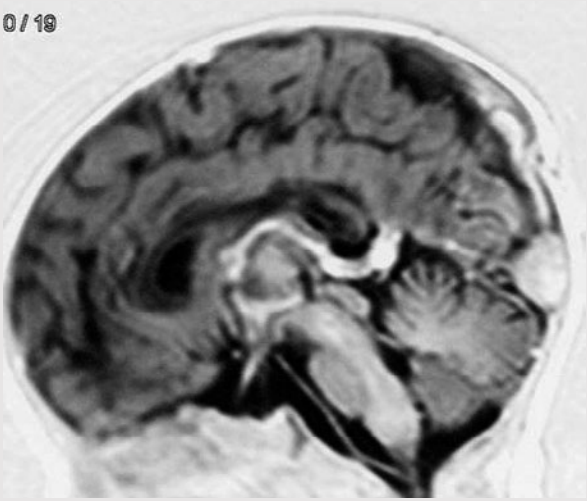


Fig. 4

Cerebral MRI, coronal view: asymmetric hemorrhagic
infarction of basal ganglia and thalamus, intraventricular bleeding and periventricular infarction on the
Cerebral MRI, sagittal view: thrombosis of internal cerebral veins, great cerebral vein, sinus rectus and superior sagittal sinus.

Fig. 5



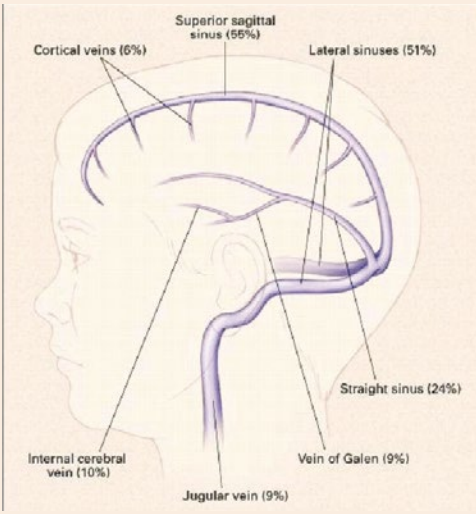
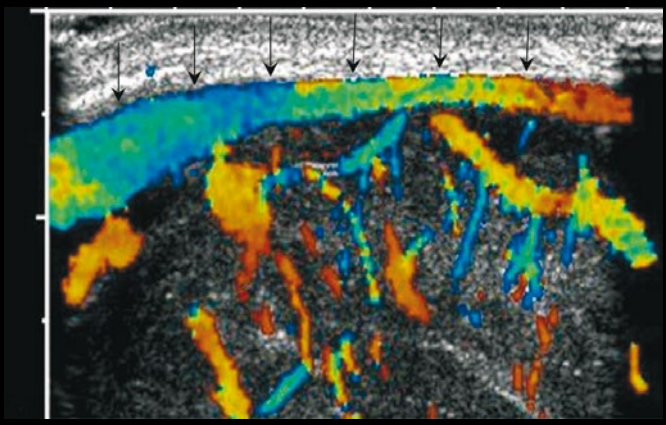


Fig. 6

Structures most susceptible to SVThr in children with the relative frequency given in parenthesis (courtesy of N Engl J Med 2001;345:420).

Normal cerebral Doppler study: superior sagittal sinus (arrows).

Fig. 7



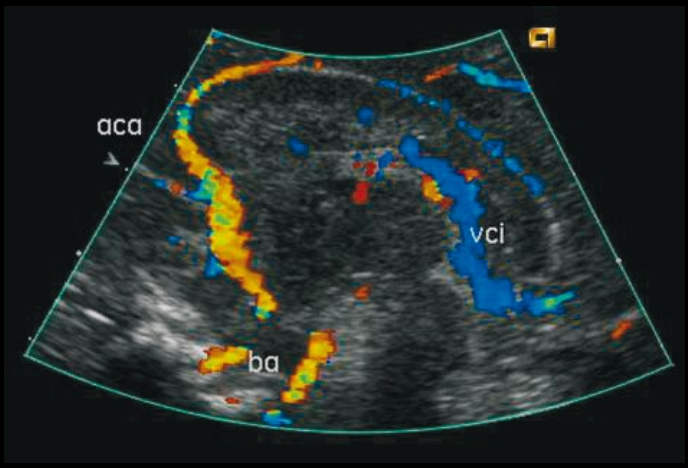
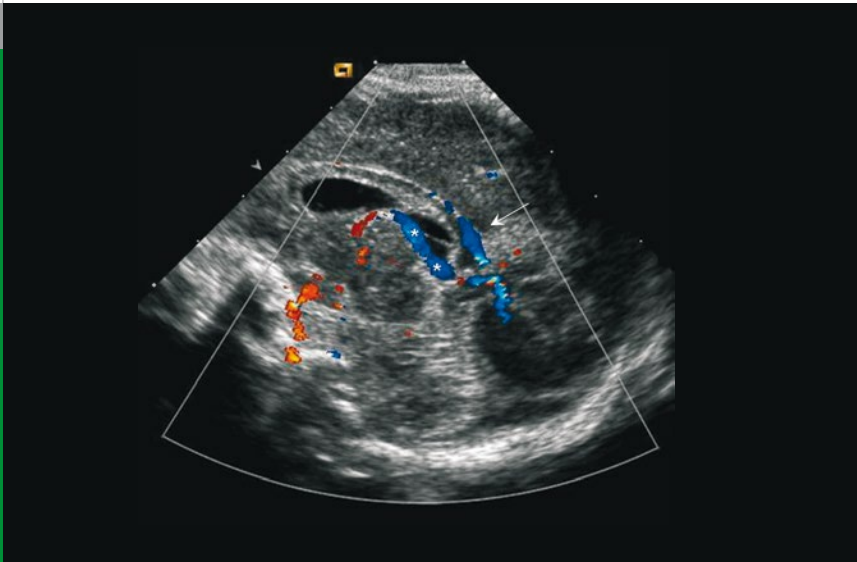


Fig. 8

Normal cerebral Doppler study: internal cerebral vein

Normal cerebral Doppler study: inferior sagittal sinus (arrow), internal cerebral vein (asterisks).

Fig. 9



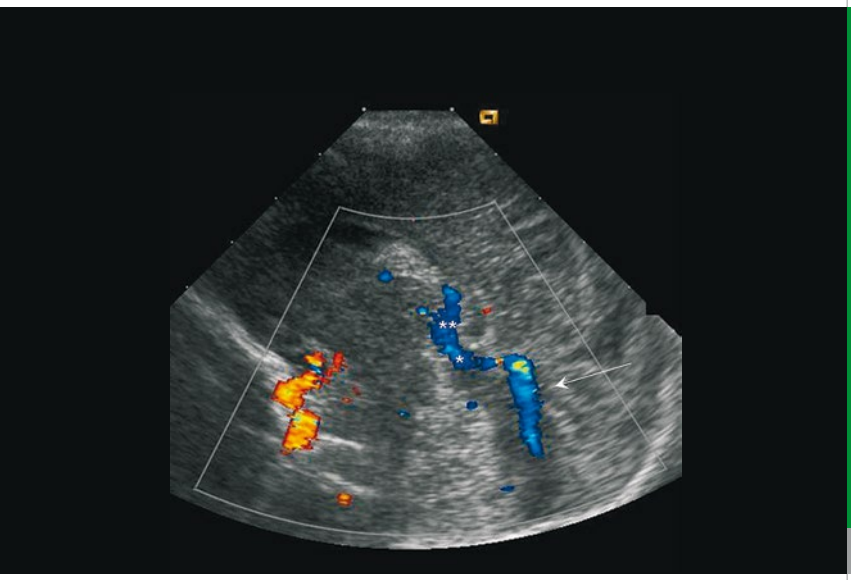


Fig. 10

Normal cerebral Doppler study: internal cerebral vein (double asterisk), vein of Galen (asterisk), straight sinus (arrow).

Normal cerebral Doppler study: straight sinus (arrow).

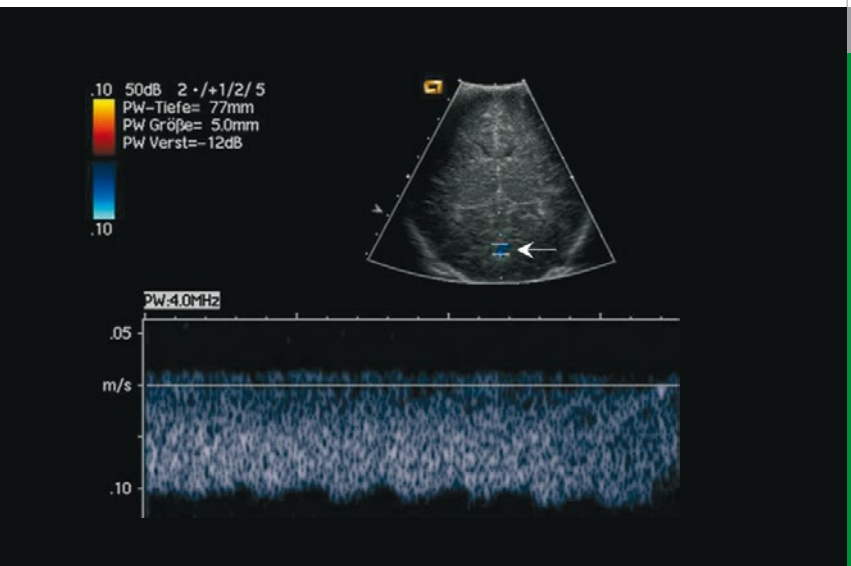


Fig. 11

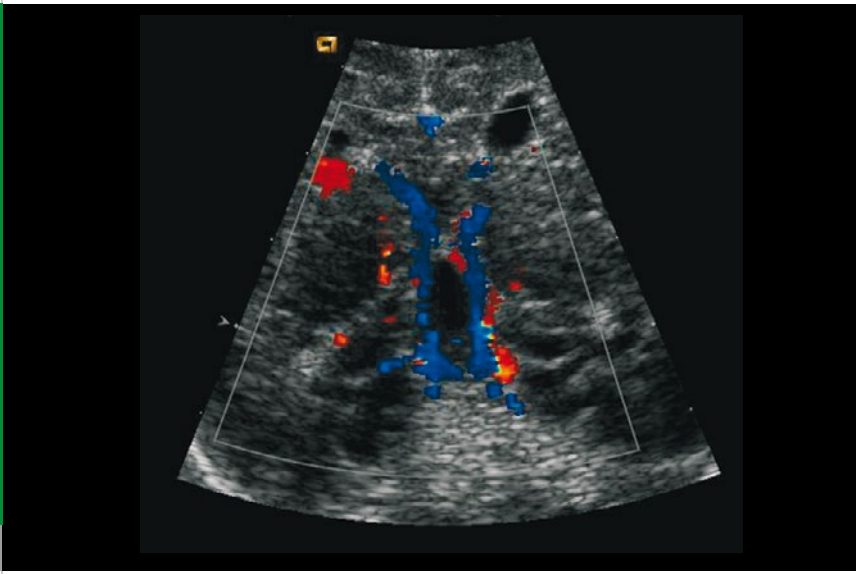


Fig. 12

Normal cerebral Doppler study: internal cerebral veins (blue) seen on occipital coronal view.

older children. DeVeber (1) observed, that seizures were seen more often in neonates (71% versus 48%) and were the most common neurologic manifestation overall. Both focal (53% versus 29%) and diffuse (90% versus 58%) neurological signs (i.e. decreased level of consciousness, headache, jittery movements, papilledema, hemiparesis, visual impairment, cranial nerve palsy) were observed more often in non-neonates. Risk factors for the development of SVThr were present in 98.6% of the neonates, the most important of which was an acute systemic illness (84%), e.g., dehydration, bacterial sepsis or pe-

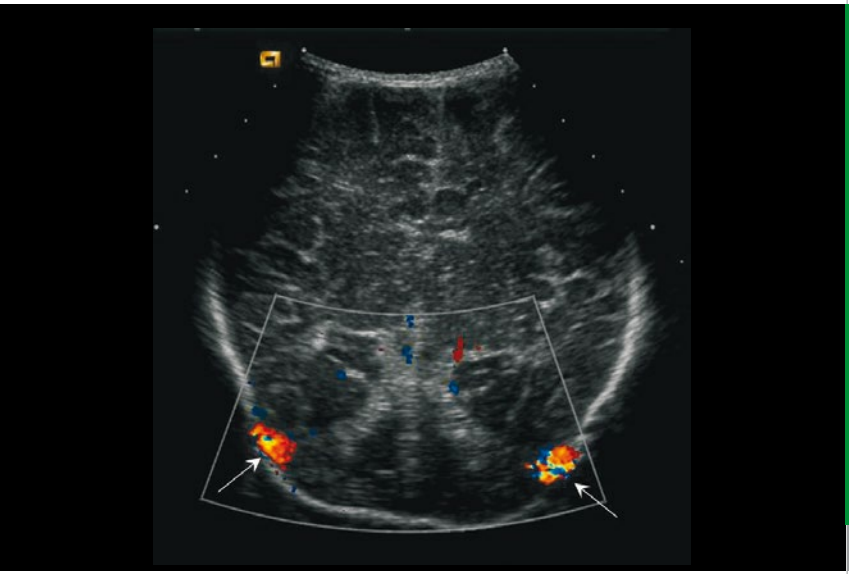


Fig. 13

Normal cerebral Doppler study: transverse sinuses (arrows).

rinatal complications (hypoxia at birth, premature rupture of membranes, maternal infection, placental abruption, gestational diabetes). In contrast, chronic systemic diseases, head and neck disorders (i.e. infection) or hypercoagulable states were seen more often in non-neonates.

Hemorrhagic infarcts as observed in our patient were found in 35% of neonates compared to 23% of non-neonates. Predictors of adverse neurologic outcomes included seizures at presentation in non-neonates and the presence of infarcts in both age

groups. The long-term neurologic outcome of SVThr in children is unclear. After a mean follow-up time of 2.1 years, deVeber found that 77% of neonates were neurologically intact (compared with 52% of non-neonates) (3).

Kenet et al (4) studied the prevalence of prothrombotic risk factors in children with SVThr in a multi-center, case-control study. At least one prothrombotic risk factor was detected in 5 of 8 (63%) neonates with SVThr, in 16 of 38 (42%) non-neonates compared to 41 of 112 (37%) children in the control group. The cause of the SVThr in our patient remains unclear. Although he refused to eat, he showed no signs of dehydration when the SVThr occurred. The only prothrombotic risk factor is his heterozygous factor V mutation.

Doppler ultrasound examination is an uncomplicated, non-invasive and inexpensive method to diagnose SVThr (for normal anatomy and normal Doppler US images see Fig. 6-13). SVThr leads to occlusion of the great cerebral vein and internal cerebral veins and, through compression of adjacent structures, can lead to hemorrhagic infarction of the basal ganglia and the thalamus which may include the internal capsule. On ultrasound images, venous hemorrhagic lesions have an irregular border and are typically bilateral, asymmetrical and often occur with intraventricular bleeding. In contrast, arterial infarcts have a regu-

lar border and typically do not involve the internal capsule and do not cause intraventricular bleeding. Nowak-Göttl et al. (5) have published recommendations for the therapy of thromboses in childhood with low molecular weight heparin. The duration of therapy depends on the presence or absence of any underlying coagulopathies and must be individualized.

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CONTACT

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