cysts are similar to those of CSF. Dermoid cysts, however, exhibit negative absorption values markedly below CSF, spread along the base of the skull and tend to be located in the midline, whereas epidermoid cysts usually occur in lateral positions.[4] Intraventricular locations in both types of cyst have been noted[7] sometimes leading to ventricular obstruction.[5] Both tumours may rupture spilling fatty components into the subarachnoid space or the ventricular system.[2] Because of their negative absorption values, fat droplets are easily detected by CT, although initially they may be misinterpreted as intraventricular air.[2] Clinical symptoms include seizures, raised ICP or aseptic meningitis[1] which may occur after spontaneous rupture or postoperatively.[2,5] Chambers[5] described a patient with communicating hydrocephalus requiring a shunt. In contrast to our patient, a solid tumour mass was identified by either CT, encephalography or histological investigation in all cases previously reported. In our case, we suggest that the intraventricular fat was derived from a ruptured dermoid cyst because of the negative absorption values (−60HU) and the location of residual fat deposits near the midline (upper quadrigeminal cistern). The occlusion of the aqueduct of Sylvius was confirmed in the sagittal MR images. We suggest that it was due to a granulomatous reaction caused by the lipid material in the neighbourhood of the aqueduct. The clinical symptoms of our patient included severe headaches, vomiting and, as a sign of raised intracranial pressure, a slowed heart rate. The rapid development of symptoms required a temporary external drainage of the lateral ventricles and followed by implantation of a ventriculoperitoneal shunt. To our knowledge, this is the first case reported where an occlusion hydrocephalus was caused by free intraventricular fatty material possibly derived from a ruptured dermoid cyst.

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Cerebral venous thrombosis in hereditary protein C deficiency

Sir: Protein C is a vitamin K-dependent serine protease which together with its cofactor protein S and antithrombin III is probably the most important inhibitor of plasma coagulation.[1-3] It is a plasma zymogen that is activated by thrombin coupled with an endothelial cofactor named thrombomodulin and inactivated by a specific plasma protease inhibitor. The anticoagulant effects of protein C are achieved through cleavage of factors Va and VIIIa of the intrinsic pathway. Clot lysis results from interaction of protein C with the inhibitor of plasminogen activator.[4]

Homozygous protein C deficiency has low or even undetectable antigenic levels and is mostly lethal through massive venous thrombosis during the neonatal period.[5,6] The heterozygous form is an autosomal dominant disorder with an incidence of 1 per 16,000 individuals. It usually presents during early adulthood as recurrent superficial and deep thrombosis of pelvic and leg veins sometimes with subsequent pulmonary embolism in several members of a given family.[7]

Diagnosis is made by repeated measurements of decreased protein C levels, while all other vitamin K-dependent factors are normal and an acquired protein C deficiency is excluded.[8] The latter occurs in liver diseases, where protein C can already be lowered yet with a normal thromboplastin time. An acquired deficiency is also observed in dissiminated intra-vascular coagulation, during coumarin treatment, and in the postoperative period.[9]

We observed a 30 year old housewife with common migraine since age 18. Since then she had been smoking five cigarettes a day and had been regularly taking an oral contraceptive. At age 25 she had suffered from two episodes of pulmonary embolism which had been treated by coumarin for several months.

In March 1986 she had sudden prickling headache bitemporally, recurrent vomiting and blurred vision. Two days later numbness of the left arm occurred, and memory deficits were noted.

On admission the obese patient had left homonymous hemianopia, a weak left arm, and a moderate paresis of the left leg. Deep tendon reflexes were brisk, moderately elevated on the left. The patient had a slight meningism, she was drowsy with diminuition of concentration and attention, but fully orientated. Routine blood examination revealed a slightly elevated activity of Gamma-GT (53 U/L). Serologic search for infectious and autoimmune disorder was unproductive. Cerebrospinal fluid was clear with a protein content of 0·38 g/l. There were two cells/mm^3 in the cell count; cell sedimentation showed few leucocytes and monocytes, but no erythrocytes or lymphocytes. CT on admission revealed abnormally increased density along the superior and the straight sinus which showed a slight enhancement after contrast medium application. There was no empty triangle sign. Patchy hyperdense areas in both parietal areas and right occipitally were interpreted as either congestioned blood vessels or petechial haemorrhages. Subsequent carotid angiography revealed lack of filling of the sinuses mentioned above with dilatation of some collateral cortical veins, thus confirming sinovenous thrombosis.

Intravenous heparin treatment was started at a daily dose of 24·000 U. Additionally sorbitol and dexamethason were given.

In spite of this treatment somnolence increased, vision in the previously intact right hemianopic fields blurred, there was an intermittently dilatated right pupil, and a choked right optic disc. Two days later short-lasting focal motor seizures of distal left arm were observed. EEG showed a moderate diffuse abnormality and a delta focus in the right temporo-occipital region with recurrent focal sharp-slow-waves.

CT four days after admission revealed a patchy haemorrhagic infarction in the right parieto-occipital region with some small haemorrhages in both parietal areas. The latter were thought to be due to anticoagulant medication, and heparin treatment was immediately stopped. From the sixth day on the patient's condition gradually improved. She intermittently reported formed hallucinations in the left

hemianopic fields, which were not associated with focal discharges in the EGG.

The patient recovered well, and up to now she has been suffering from infrequent sensory Jacksonian seizures of the left arm which are treated with oral carbamazepine. Residual findings are a right optic nerve atrophy and slightly elevated tendon jerks of the left arm.

Several members of the patient's family had a history with thrombosis and pulmonary embolism. Coagulation studies in all these probands revealed normal values of partial thromboplastin time, thromboplastin time, thrombin time, factors I, II, V, VII, VIII, IX, X and of antithrombin III. Protein C levels were immunologically determined by an ELISA test.

Functional measurement was done by an amidolytic test with a chromogene substrate determined photometrically and by a coagulation test with inhibition of activated partial thromboplastin time.[10]

The table indicates that protein C values of the index patient were subnormal in the ELISA, normal in the amidolytic test, and lowered in the coagulation assay. Several family members had more markedly decreased protein C levels with less or even no clinical involvement.

After obtaining these results oral phenprocoumon administration was started in the patient 3 weeks after admission. During the first 2 weeks of this therapy again high-dose heparin was given. The patient is now under out-patient care without any complications of thromboembolism or of the combined anticonvulsant and anticoagulant medication.

Heterozygous protein C deficiency is an important predisposing risk factor for thromboembolic disease in young adults.[2 7] Neurological abnormalities due to cerebral sinovenous thrombosis seem to be rare in this condition. So far five patients have been sufficiently documented in the literature.[11-13] In one study, three female patients had prodromal headache, seizures, hemiparesis, and altered mental status. CTs in two of these revealed haemorrhagic infarction and the angiogram in one patient showed a superior sinus thrombosis. In one patient a haemorrhagic infarction in the basal ganglia was seen at necropsy.[11]

In a case report, a female patient had hemiparesis, aphasia, and seizures. CT showed the empty triangle sign; superior and inferior sinus thrombosis was seen in the angiogram.[12] The fifth patient, a 32 year old male, had an angiographically demonstrated "cerebral thrombophlebitis".[13]

All reported patients with CNS involvement had at least one relative with a lowered functional and/or antigenic protein C level and/or with a history of thromboembolic disease elsewhere in the body. As is shown by our family data too (table) heterozygous protein C deficiency itself does not inevitably cause clinical disease. Several family members had far more decreased protein C levels than the index patient, but were not or only slightly affected clinically.

Table *Clinical presentation and protein C values in the pedigree of the index patient* (DVT = deep venous thrombosis; PE = pulmonary embolism).*

Patient	*Age (yr)*	*Presentation*	*ELISA (70–140%)*	*Amidolytic (70–140%)*	*aPTT inhibition (60–150%)*
Father	62	DVT, PE	40	39	29
Mother	65	—	85	117	96
Sister	36	Cigarettes, oral contraceptive	34	36	22
Niece	4	—	66	61	50
Nephew	2	—	35	33	20
Sister	33	DVT, PE, oral contraceptive	45	45	29
Index case*	30	DVT, PE, oral contraceptive, sinovenous thrombosis	63	73	41
Sister	25	DVT, oral contraceptive	44	43	26

It is therefore suggested that additional thromboembolic risk factors, such as cigarette smoking, oral contraceptives, obesity, pregnancy, puerperium, trauma, or surgery considerably contribute to the clinical manifestation of thrombosis also in the cerebral sinus.[14] In our index patient several of these factors were present. In one of the other neurologically affected patients previous oral contraception has been reported.[11]

In contrast to cerebral sinovenous thrombosis there is no association of protein C deficiency to thrombosis of cerebral arteries.[15 16] In one report, diminished protein C levels in acute stroke were still in the normal range and were related to the haemostatic derangement caused by the event itself, but did not point to a hereditary condition.[17]

In our patient the clinical and radiological investigations indicated that her cerebral haemorrhagic infarction was due to sinovenous thrombosis, the occurrence of which was related to the hypercoagulable state present in protein C deficiency. In the past, anticoagulant medication in acute sinovenous thrombosis has been controversial. The potential benefit of heparin to prevent the development and propagation of thrombus has to be weighed against the possibility of promoting intracerebral haemorrhage.[18 19]

According to the literature and our own experience anticoagulant therapy nowadays not only provides thrombolysis as demonstrated by angiography.[20] It does not cause deterioration of the clinical condition and outcome, even when intracerebral haemorrhages have been detected prior to treatment.[21] In our patient high-dose heparin administration was stopped, when haemorrhages were observed under therapy, which had not been found by the first CT.

In the acute stage of a thromboembolic event due to protein C deficiency heparin anticoagulation is strongly advised.[22] The prophylactic administration of coumarin is recommended after one or more thromboembolic events; on the other hand, children with the heterozygous form of the disorder and adults, who are not affected clinically, do not require anticoagulative treatment.[14 22] In our case coumarin prophylaxis was started for the following reasons: (1) Cerebral sinovenous thrombosis is a much more life-threatening condition than thrombosis of extremities. (2) The primary coagulation defect persists even after survival of the acute disease and thus offers a high risk of relapse, especially when clot lysis was incomplete or when the persisting thrombus is not yet coated by an endothelial layer.

During the initial coumarin treatment period high-dose heparin administration must be sustained, because the protein C concentration falls dramatically with coumarin intake due to the short half-life period of protein C (6 hours). Simultaneously the inhibition of synthesis of factors II, VII, IX, and X is still delayed. Heparin therefore prevents enhanced clotting which during this period may rarely result in a haemorrhagic necrosis of the skin, and up to now poorly understood complication.[23]

Thus, in cerebral sinovenous thrombosis of young patients with a personal or family history of recurrent venous thrombosis elsewhere in the body, and with lowered protein C levels, a lifelong treatment with vitamin K antagonists does seem to be advisable.

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Geometrical optics of the retinal image stabilisation device

Sir: An optical device giving partial or almost-complete stabilisation of the retinal image has been described[1] and tested[2 3] to treat patients with continuous oscillopsia caused by nystagmus, usually due to acquired neurological disease such as multiple sclerosis. It can relieve such oscillopsia, and consists of a high-plus spectacle lens used together with a high-minus contact lens.

In many of these patients the amplitude of oscillopsia is less than would be predicted from the amplitude of their nystagmus, so that the image only needs to be partially stabilised. A means is then needed for predicting how much retinal image stabilisation ("RIS") will be needed in each case, and for calculating the correct lens power required to achieve it.

An index of oscillopsia has been developed, defined as the ratio of angular amplitude of oscillopsia ("O") and angular amplitude of nystagmus ("N"). O/N can vary from 0 (no oscillopsia, in spite of nystagmus) to 1 (oscillopsia of the same amplitude as nystagmus). The index of retinal image stabilisation required is also equal to O/N, since the proportion of the nystagmus that is perceived as oscillopsia is also the proportion of the nystagmus that must be cancelled by the device.

Partially stabilising the retinal image is equivalent to increasing the amount of rotation of the eye required to move the image through a given angle on the retina (rotational magnification). Retinal image stabilisation therefore bears a simple relation with rotational magnification, which is a well-studied property of plus spectacles.

Rotational magnification of the image on the retina (RMr) is here defined as the ratio of the angular motion of the eye scanning a scene (N) and the resulting angle of retinal image-slip (P). RMr differs from the amount of rotational magnification in terms of distance across the scene (RMs) when, as here, the device causing the rotational magnification also causes angular magnification of the retinal image. What we need is to apply sufficient RMr to reduce the retinal image-slip (originally equal to N) by the amount of the oscillopsia (O). The remaining image-slip P does not cause oscillopsia since it is compensated by a central mechanism ("C"), whose site and mode of action is not known.

Then if:
N = amplitude of nystagmus
O = amplitude of oscillopsia
C = amplitude of central compensating signal
P = amplitude of residual retinal image-slip
N = C+O (1)
RMr = N/P (2)
From (1), O/N = 1−(C/N)
From (2), (RMr−1)/RMr = 1− (P/N)

For optical neutralisation of oscillopsia, we have said that P = C
So O/N = (RMr−1)/RMr = RIS (3)

The RMr is generated by the combination of spectacle and contact lens, which if the image is in focus on the retina form a galilean pair. A galilean pair is afocal, so where:
fs = focal length of spectacle (in metres)
Ps = power of spectacle (in dioptres)
fc = focal length of contact lens (in metres)
Pc = power of contact lens (in dioptres)
d = separation between principal planes of spectacle and contact lens (in metres)
fs = −fc+d
Or, 1/Ps = d − (1/Pc) (4)

The galilean pair also causes angular magnification of the image (AM), which is given by:
AM = −Pc/Ps (5) (see ref 4, p205).